



REVIEW / *Musculoskeletal imaging*

Tumours and pseudotumours of the soft tissue in adults: Perspectives and current role of sonography

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KEYWORDS

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Elastography

Abstract Soft tissue tumours of the musculoskeletal system are reported relatively frequently. The quality of the information gained from different imaging modalities (Doppler sonography, multislice CT, MRI spectroscopy, and diffusion MRI) means that in a growing number of situations, we can envisage determining with great accuracy not only the usual information of tumour size and topography, but often the exact nature of the tissue, almost always identifying whether a lesion is aggressive or not. Of all these techniques, Doppler sonography has become the most widely used due to the striking improvements in its sensors, especially for superficial applications. Some other recent developments are: panoramic imaging, elastography (although its current contribution is still to be determined but it seems to offer promising potential), and, most importantly, specific contrast agents. These techniques have considerably refined the quality of the information obtained, and have particularly enhanced the degree of sensitivity with which lesion progression can be assessed. Ultrasonography is the very first investigation in our protocol. It is also very often used to close investigations, as it accurately guides core needle biopsy from these generally accessible lesions. The purpose of this article is to bring together updated information on the various collections of sonographic features seen in soft tissue tumours and pseudotumours and to emphasise the considerable contributions of these new technological developments, in particular contrast-enhanced sonography. The discussion will follow the World Health Organisation's anatomical pathology classifications of soft tissue tumours. We will close with a synthesis that summarises the main steps in our diagnostic process.

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The spectacular improvements that have been made in superficial ultrasound imaging mean that sonography currently has an important role in the exploration of soft tissue tumours and pseudotumours. It should open the imaging investigations [1]. Apart from its contribution in terms of positive diagnosis, recent technical advances (specific contrast agent, elastography) have opened up interesting possibilities in terms of lesion characterisation.

Technique

We use an Aplio XG system, model SSA-790A, from the Toshiba Medical Systems Corporation (Zilverstraat 1. 2718 RP, Zootermeer, Netherlands). We always begin the examination using a high-frequency linear electronic transducer (8–15 Mhz). The multiple planes give us an overall view of the lesion and allow it to be measured in three spatial dimensions. When the mass is large, we take panoramic views that generally provide us with an exhaustive image of it that depicts all of its margins. For larger patients or in some anatomical areas (proximal thigh, buttocks), differently shaped transducers are required (curved surface) and a lower frequency (4–8 Mhz) is needed [2].

We also always use the different types of Doppler sonography (power, color and pulsed). Contrast-enhanced sonography forms an integral part of our protocol. We use an agent with a low mechanical index, consisting of an inert gas (sulphur hexafluoruride) stabilised by a fatty acid shell. We inject an ampoule of this as a bolus “pushed” by an infusion of normal saline solution at maximum output. It only diffuses into the vascular system. Contrast uptake, identified using VRI technology (vascular recognition imaging, which visualises both tissue, using normal frequency imaging, and vascularisation, using broadband Doppler imaging), is displayed as nodule-shaped areas that are either red or blue, depending on their orientation in relation to the transducer [3].

Elastography is still in its infancy in this field, but several studies have recently allowed us to anticipate that it will have real value for musculoskeletal imaging applications [4,5]. It should, in the near future, develop a more important role because of the valuable information it provides on the composition of lesion tissue (cellularity, extent of fibrosis) in comparison to adjacent healthy tissue.

For now, there is no proof that 3D imaging is of significant interest because of insufficient image quality. The improvements to transducers should relatively quickly allow it to play a role in these types of investigations because it offers the potential for a high quality frontal view. “Cross beam” techniques, or, in other words, orientations from the variable angles of the ultrasound beam, in our opinion do not notably improve the analysis in terms of either the content or margins of the lesion.

Sonographic analysis of lesions

Naturally, lesions will be assessed in view of the basic clinical features: speed of tumour growth, existence of pain, age and sex of the patient, and topography of the tumour. It is very useful to have standard radiographs available before

carrying out sonography. They can provide valuable diagnostic information [4]: radiolucent clarity of a fatty lesion, calcifications with fine outlines (phleboliths) in a haemangioma, linear or crescent-shaped calcifications in myositis ossificans, or even bone changes, which can be the consequence or origin of a tissue abnormality.

Sonography attempts to accurately gauge lesion size, and this process requires greater delicacy the larger the lesion. Equally, precisely situating the tumour’s topography can prove to be difficult when there is extended involvement in one compartment, or indeed to a greater extent, when multiple compartments are affected. There are numerous parameters to be taken into account when addressing the nature of a tumour or at least distinguishing an aggressive lesion from an inactive process [5]: is there a capsule, is it regular or otherwise, what is the echostructure, is it composed of tissue or fluid, and is it homogenous? It is crucial to look for zones of necrosis, calcifications, and fine or thick septations, and to assess whether the lesion is connected to a vascular, neural, or joint structure [6,7].

Doppler imaging demonstrates hypervascularisation that may be regular or anarchic (are there loops, areas of stenosis, occlusions, unbalanced or irregular vascular branches?) while pulsed wave Doppler may show localised accelerations in flow (stenoses) or a low resistive index (arteriovenous shunt) [8,9]. Does the administration of an ultrasound-enhancing contrast agent lead to enhancement? Is this early, prolonged, fleeting, or late? This contrast enhancement, in all its different manifestations, is usually a sign of an aggressive lesion, as was shown in a preliminary study on 80 cases [10], with, however, some particularities for specific tumor types (desmoid tumors). If the lesion no longer enhances further to treatment, this seems to be an argument for a favourable prognosis, at least in some types of tumours [3].

All of these parameters form an initial evaluation, which in a number of cases will lead to diagnosis: (lipoma, synovial cyst, ganglion cyst, vascular malformation, abscess, haematoma etc.) [6]. It often allows the distinction to be made between benign and malignant lesions in view of the collection of signs that are suspicious: large lesion size, usually with indistinct margins, anarchic vascularisation seen on power Doppler (Fig. 1) (occlusions, arterial stenosis, arteriovenous shunts), zones of necrosis (Fig. 2), a pattern of

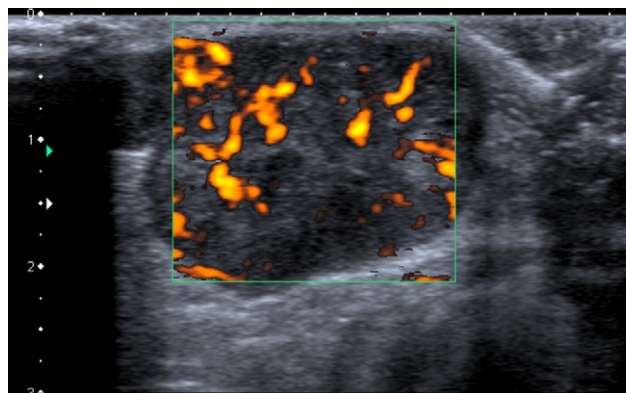


Figure 1. Power Doppler showing anarchic vascularisation (stenoses, anastomoses, “unbalanced” vascular branches) fitting a picture of an aggressive lesion.

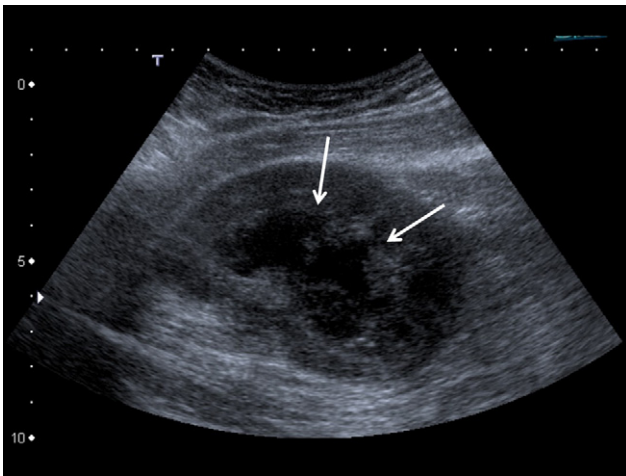


Figure 2. 75-year-old male. Investigation for a fast progressing soft tissue mass of the thigh. Sonography shows central zones of necrosis (white arrows) within a large solid mass, suggestive of an aggressive lesion.

spread across the aponeuroses, and early and significant contrast uptake (Fig. 3) are all features suggestive of an aggressive lesion [9]. This must, however, be contextualized: arteriovenous shunts are also seen in vascular malformations, and irregular tumour margins are sometimes seen in truly benign lesions, as is striking contrast uptake (some desmoid tumours).

This is why, except in some rare cases when sonography does lead to a confirmed diagnosis, complementary investigations are usually required. These include principally MRI, due to its accuracy in assessing topography and size, and its ability to determine lesion characteristics, and CT, which best identifies bone changes as well as calcifications in the soft tissue [11,12]. Nonetheless, there is a return to sonography to close diagnostic investigations by means of ultrasound-guided core needle biopsy of the zones within the tumour that have the potential to be the most informative (Fig. 4) [13].

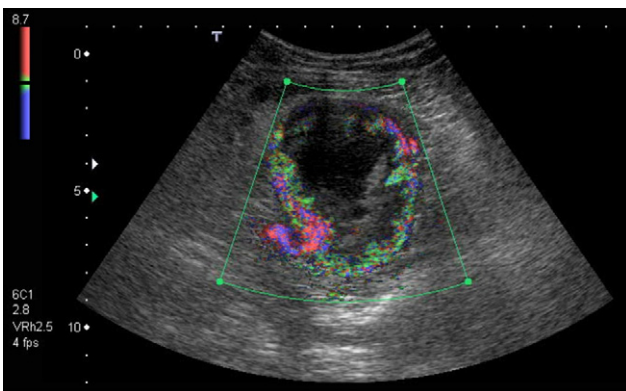


Figure 3. In the same patient, a quick (20 seconds) and considerable uptake of sonographic contrast agent. A strong argument in favour of an aggressive lesion (images 4 and 5 are drawn from a report of a dedifferentiated liposarcoma).



Figure 4. Ultrasound-guided needle biopsy of a soft tissue "tumefaction" of the arm. Needle clearly visible (white arrow).

Lesion classification based on anatomical pathology

For the purpose of clarity of the discussion, we must lean on a framework that is commonly accepted internationally. As a guiding principle we will use the World Health Organisation's (WHO) anatomical pathology classification of soft tissue tumours [4]. It describes ten groups of soft tissue tumours:

- adipocytic tumours;
- fibroblastic/myofibroblastic tumours;
- fibro-histiocytic tumours;
- smooth muscle tumours;
- perivascular tumours;
- skeletal muscle tumours;
- vascular tumours;
- chondro-osseous tumours;
- neurogenic tumours;
- tumours of uncertain differentiation.

There are more than 80 possible histopathological diagnoses, but a small number of lesion types account for the vast majority of cases (at least 80 percent), and these include both benign and malignant lesions. (The relative proportion of malignant to benign lesions is one in one hundred).

We will firstly describe those pseudotumours with well-established sonographic signs: synovial cysts, ganglion cysts, haematoma, abscess, aneurysms and pseudoaneurysms, and epidermoid cysts. Even though these are not true instances of neoplasm, it is useful to have a good understanding of their sonographic appearance because they are common and need to be distinguished from true discrete masses.

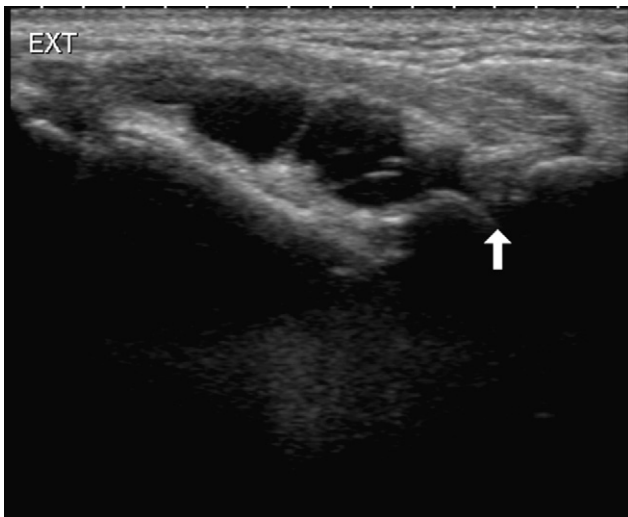


Figure 5. Tender tumefaction on the dorsal surface of the wrist in a 52-year-old woman. Sonography shows a cystic formation with septations that is “connected” to the joint (white arrow) suggesting a synovial cyst.

Commonly encountered pseudotumours

Synovial cyst

This is a “diverticulum” of the synovium that lines the joints, which is fluid-filled, can contain several echogenic zones, sometimes has septations, and varies in size from minimal to significant. It is crucial to demonstrate the essential feature of diagnosis: that there is continuity between the cyst and the joint (Fig. 5). This common lesion is readily found in two locations in our experience: the wrist, usually the dorsal surface, issuing from the interline separating the radial and ulnar epiphyses and the first row of the carpal bones; and the knee, developing within the semimembranosus bursa and medial head of the gastrocnemius muscle (Fig. 6) It can be large and cause disability (meaning ultrasound-guided drainage is indicated), or it can rupture (pseudothrombophlebitic picture) [13,14]. Any other joint can be affected, but this is less common. The

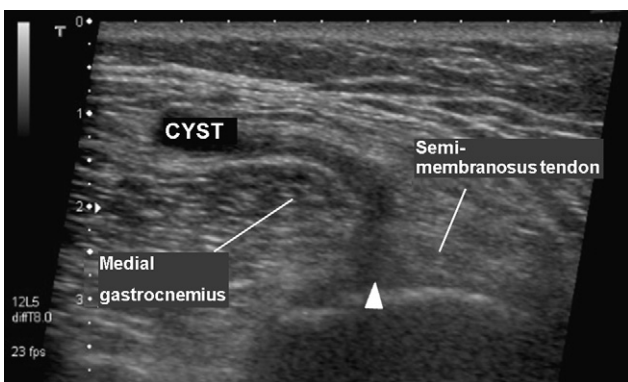


Figure 6. Popliteal cyst: topographical landmarks. Tumefaction of the popliteal fossa in a 56-year-old male. Axial view of the popliteal fossa highlights the cyst between the semimembranosus muscle and the medial head of the gastrocnemius muscle. Communication (arrowhead) with the joint.

signs on sonography would be the same as those described above.

Ganglion cysts

These are named for their morphology, and they are adherent to the joints (although communication has sometimes been lost), tendon sheaths, or bursae. These formations are made up of concentrated synovial and mucoid fluid [2,6,13]. They are lined with non-contiguous, flattened synovial cells and connective tissue. Sometimes large, their impact on the adjacent vessel and nerve structures can translate into clinical signs (disability, pain, paraesthesia etc.). There are some localisations that are classically seen: the dorsal surface of the wrist close to the scapholunate ligament; adjacent the spinoglenoid notch (Fig. 7a) on the posterior surface of the shoulder complicating a labral fissure and sometimes involving the suprascapular nerve; on the hip, as a result of the same process; and on the knee, due to a meniscal lesion. They can also develop in contact with a nerve or an artery (Fig. 7b) [15].

Haematoma

These often appear in a suggestive context (direct or indirect trauma in athletes) or in elderly people being treated with vitamin K antagonists. They have an echostructure that changes over time: although relatively echogenic at first, the contents gradually become almost entirely liquid (Fig. 8), usually starting at the periphery and progressing towards the centre, and in the final stages the centre alone is slightly echogenic. They are avascular. They can persist, becoming calcified at the periphery, or continuing to bleed (chronic haematoma). Any haematoma of the soft tissue that does not fit into this classic pattern, and in which the circumstances causing it are not clear, must be considered with suspicion and be subject to close monitoring. Haematoma can accompany or “mask” a true neoplastic lesion. MRI and sometimes a biopsy [2,6] will be necessary.

Abscess

These can be seen in a context of general infection or secondary to local “traumas” (injections in drug users), and are facilitated by a state of immunodeficiency. The content is usually fluid, often with a few areas producing low amplitude internal echoes, and they are sometimes scattered with clearly visible septa. They are enclosed in a thick capsule (Fig. 9) and are avascular. Sonography is useful to guide punctures for bacteriology sampling and drainage [2,6].

Vascular pseudotumours

These may be true aneurysms (often in the popliteal fossa) that are generally fusiform, with the edges of the arteries losing their parallel structure, and they can sometimes be part of a syndrome in which multiple aneurysms are formed. Otherwise they may be pseudoaneurysms secondary to local trauma (bone fractures, injections in drug users, instrumental manoeuvres in interventional radiology) and color Doppler imaging has made distinguishing them straightforward. It demonstrates, except in rare cases of

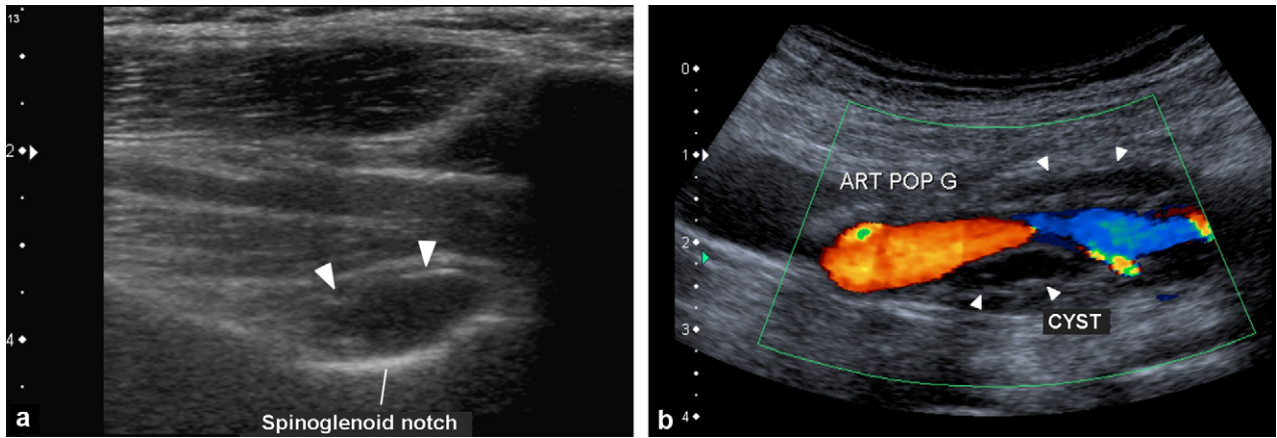


Figure 7. a: posterior axial view of the shoulder just below the scapular spine in a patient with chronic pain: presence of an ovoid formation of the spinoglenoid notch (arrowheads): ganglion cyst secondary to a labral fissure demonstrated on a CT scan of the joints; b: longitudinal view of the popliteal fossa in a 59-year-old male who had undergone an operation several years previously for a “cyst” in the region: cystic formation with septations (arrowheads) “enveloping” the popliteal artery: ganglion cyst.

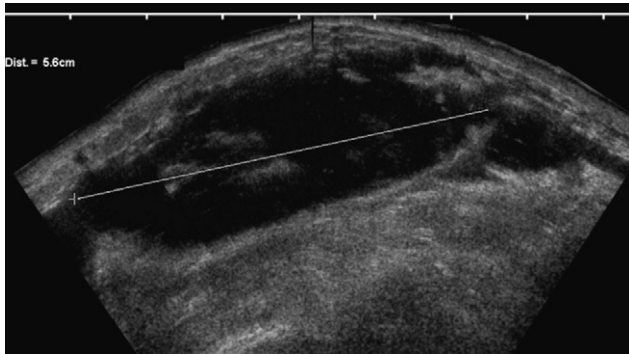


Figure 8. Longitudinal view of the thigh in an elderly patient treated with vitamin K antagonists, who had had a fall a few days previously: large fluid-filled formation corresponding to a haematoma undergoing “liquefaction”.

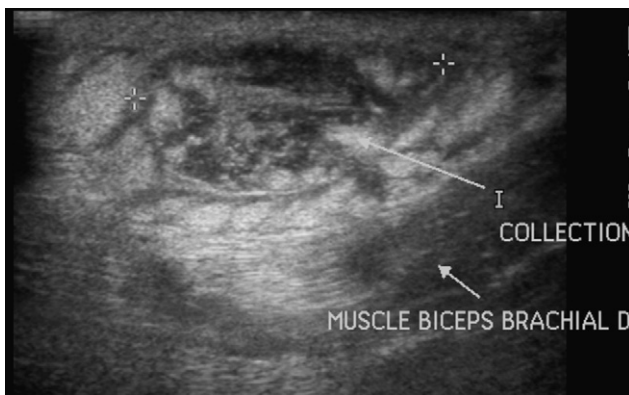


Figure 9. Fluid collection with echogenic, non-homogenous contents and a thick shell (long white arrow) which has a highly inflammatory appearance, secondary to an injection in the soft tissue of the arm in a drug-using patient: abscess of the soft tissue.

almost complete thrombosis, linear blood flow within true aneurysms, and swirling blood flow within pseudoaneurysms, in which another finding is communication between the “mass” and the arterial lumen (Fig. 10) [2,13].

Epidermoid cysts

These are found at very superficial sites and are oval with a soft tissue echostructure. They are slightly echogenic, homogenous overall, having just a few internal linear hyper-echoic areas (corresponding to keratin), and their margins are well-defined except at each extremity, where acoustic shadowing is frequently seen. No vascularisation is found and the thickness of the subcutaneous layer of fat is normally reduced in respect of the mass (Fig. 11) [6].

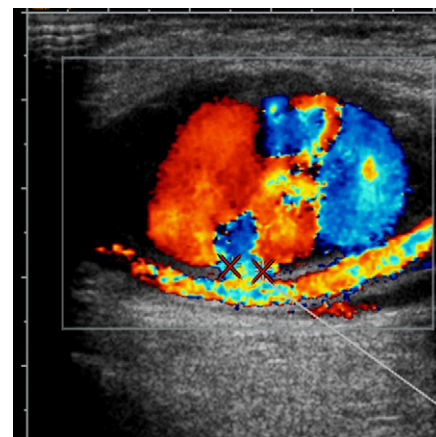


Figure 10. Color Doppler sonography showing a pseudoaneurysm of the femoral artery complicating an interventional radiology procedure. The communication between the artery and fluid collection (red crosses) is clearly visible and the flow within has a whirlpool-like appearance (flow colored red and blue).

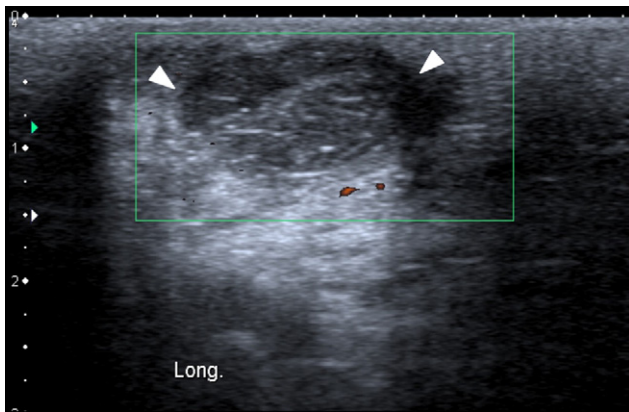


Figure 11. Sonography of the upper part of the back: Superficial, solid, ovoid formation that is non-homogenous, avascular on Doppler imaging, and contains linear echogenic zones. This is an epidermoid cyst.

Adipocytic tumours

Lipoma

Lipoma is the most common benign soft tissue tumour, and it is usually situated in the subcutaneous tissue. It produces a fusiform image with its long axis parallel to the skin line, has a soft tissue echostructure, is homogenous, being hyperechoic overall, and it has a micronodular appearance due to very fine septations with the lesion [2,5,6,14]. It has ill-defined margins (Fig. 12). Power Doppler demonstrates an absence of vascularisation. There is no enhancement after contrast injection. Other deep forms may be encountered, and these may be intra (Fig. 13) or intermuscular [16]. They usually have the same sonographic features, but “atypical” features are sometimes seen: large size, permeative margins, or heterogeneous echostructure, consisting of nodules with variable appearances or thick septations [17]. All these unusual features in a fatty lesion call for caution, and MRI

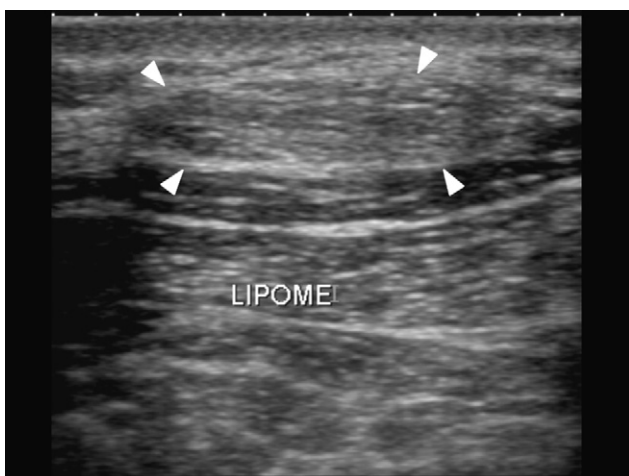


Figure 12. Sonography of the neck in an 80-year-old patient who had for a number of years been aware of a painless tumefaction that was stable in size on the posteroinferior part of the neck: hyperechoic mass sited in the subcutaneous tissue with narrow margins that is avascular (arrowheads): superficial lipoma.



Figure 13. Longitudinal view of the thigh in a 52-year-old woman who had reported a painless swelling in this area that had progressed over several years: intramuscular lipoma (arrowheads) in the anterior compartment of the thigh.

and biopsy should be carried out in order to exclude a well-differentiated liposarcoma (containing at least 75% adipose tissue), as this is the main differential diagnosis.

Malignant adipocytic tumours are often large in size, sometimes painful, and are mainly seen in men in the proximal lower limbs. The wide variety of histologic types (myxoid, round cell, pleomorphic, undifferentiated) explain why there are so many possible presentations on sonography, which have in common an aggressive appearance, with occasional calcifications. Naturally this means that further investigations and biopsy are indicated (Fig. 14a and b).

A specific case: neural fibrolipoma

This usually affects the median nerve. There is a tumefaction of the wrist that is tender on pressure, which on axial plane sonography appears as an oval formation that is centred on the nerve with multiple small hypoechoic nodules (Fig. 15). On longitudinal views it appears as multiple intercommunicating fascicles [2,5,14,18]. It has a heterogeneous appearance because it is composed of a juxtaposition of hyperechoic fatty tissue and hypoechoic nerve tissue. It is avascular on Doppler imaging, and the use of a contrast agent has no effect. MRI features overlap with the sonographic findings.

Fibroplastic/myofibroplastic tumours

Nodular fasciitis

This a benign and reactive proliferation of myofibroblasts that is usually seen in young adults at the extremities of the upper limbs, with the neck and lower limbs being the next most common sites [19]. More rarely it can develop in muscle and it is then known as proliferative myositis. On sonography it appears as a solid oval formation with low or iso echogenic that is well-defined with regular or

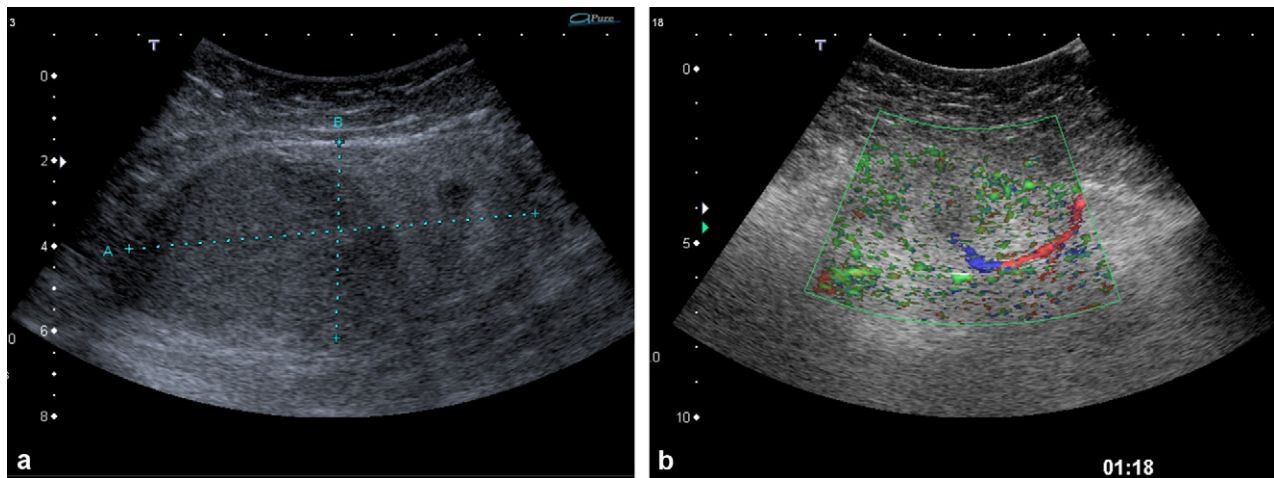


Figure 14. a: longitudinal view of the internal aspect of the thigh in a 75-year-old female who reported a tumefaction that had developed relatively recently (one year) and was growing in size: a voluminous, solid mass on the internal aspect of the thigh that is non-homogenous and consists of large hyperechoic areas (fatty tissue); b: contrast-enhanced sonography view showing contrast uptake, a strong argument in favour of an aggressive lesion. Anatomical pathology diagnosis: dedifferentiated liposarcoma.

slightly lobulated margins, and is avascular both with and without contrast material administration. Its main characteristic is that it maintains a connection with a connective tissue sheath, fascia, or aponeurosis.

Fibroma of tendon sheath

This affects adults between the ages of twenty and fifty. It manifests as a lesion similar to a giant cell tumour, and these two could be considered together as one entity. The only differences between them are variations in their respective distributions of cellular and stromal collagen content, with fibroma being low in cells and rich in connective tissue, and the opposite being true for giant cell tumours. Between these two ends of the spectrum, all possible distributions may be found. Their clinical and sonographic features are identical (see giant cell tumours) [19].

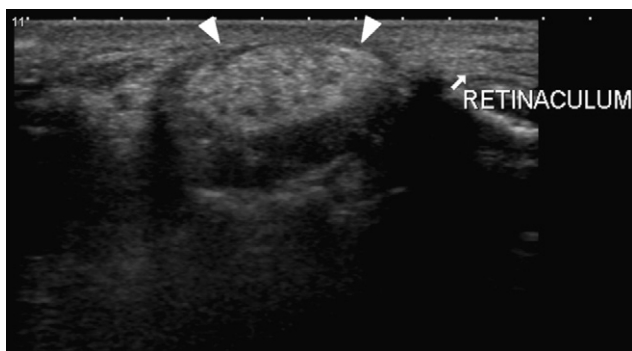


Figure 15. Axial sonographic view of the anterior aspect of the wrist in a young 25-year-old woman who reported a tender tumefaction that had developed several years previously. Multilobulated formation that is non-homogenous, with both hypoechoic and hyperechoic areas, centred on the medial nerve (arrowheads): neural fibrolipoma.

Elastofibroma

An elastofibroma is a reaction to a repeated mechanical irritation (pseudotumour) rather than a true neoplasm. It is usually situated in the connective tissue between the posterior chest wall and the inferior scapular angle, deeper than the rhomboid muscles and latissimus dorsi muscle in relatively elderly patients (mean age: 70). Other localisations are possible: the peri-trochanteric area of the hip, or in the elbow adjacent to the olecranon. It is sometimes bilateral. Clinically, there is pain and induration. On histology, it is made up of an accumulation of collagen and abnormal elastic fibres that is interspersed with a contingent of cells, adipose tissue, fibroblasts, and myoblasts. The image seen on sonography is one of a well-defined, ovoid formation with its long axis parallel to the wall. It is made up of tissue, and it is heterogeneous, having striations alternating with linear or curved bands that are hyper and hypoechoic (Fig. 16). It shows no enhancement after contrast injection [20,21].

Superficial fibromatosis

These originate in the palmar or plantar fascias or aponeuroses. They are made up of fusiform myofibroblastic cells, intercellular collagen deposits within a myxoid matrix and "constricted" vessels [19,22].

Palmar fibromatosis (Dupuytren's contracture) [22]

This affects the palmar aponeurosis on the anterior surface of the hand. It causes an indurated subcutaneous thickening with finger contractures. It mainly affects men over the age of thirty, with a clear predominance in the sixth decade. It is often bilateral, and appears on sonography as hypoechoic avascular thickening of the aponeurosis, most commonly affecting the fourth finger, followed by the fifth, third, and finally the second finger, without flexor tendon involvement (Fig. 17). Postoperative recurrence is common.

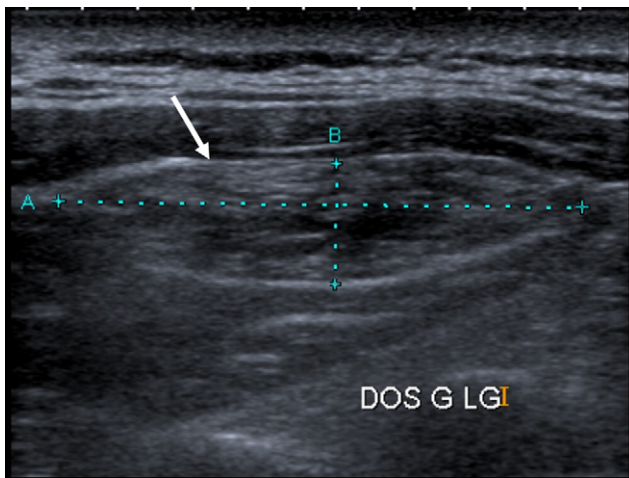


Figure 16. Transverse view of the back, at the lower internal pole of the scapula, in a 73-year-old male who reported a tender and firm tumefaction dating back several years: solid, ovoid, well-defined, deep mass that is avascular and non-homogenous, consisting of alternating hyperechoic and hypoechoic bands (white arrow): elastofibroma confirmed on biopsy.

Plantar fibromatosis (Ledderhose’s disease) [22]

This leads to single or multiple confluent nodules along the central or medial component of the plantar aponeurosis. It is sometimes bilateral. Concomitant presentation of the palmar form is possible. It affects both sexes equally, generally between the third and fifth decade. On sonography, one or several confluent nodules are visualised that are oval, solid, hypoechoic, avascular, circumscribed margins, connected into the superficial part of the plantar aponeurosis. They can spread into deep tissue, muscle, or superficial subcutaneous tissue, but this is rare. They do not enhance after contrast agent injection (Fig. 18). If symptomatic, they are

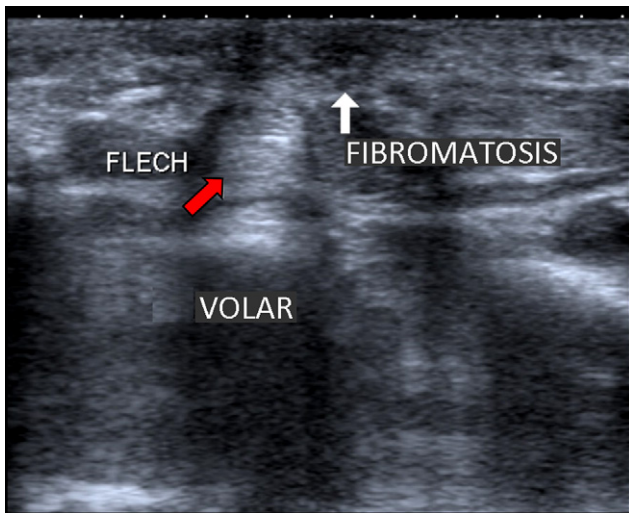


Figure 17. A 58-year-old male who reported a nodular “induration” on the volar aspect of the hand at the base of the fourth and fifth fingers with retraction and flexion of these fingers. The axial sonography view demonstrated thickening of the palmar aponeurosis (white arrow) above the flexor tendons (red arrow): palmar fibromatosis (Dupuytren’s contracture).

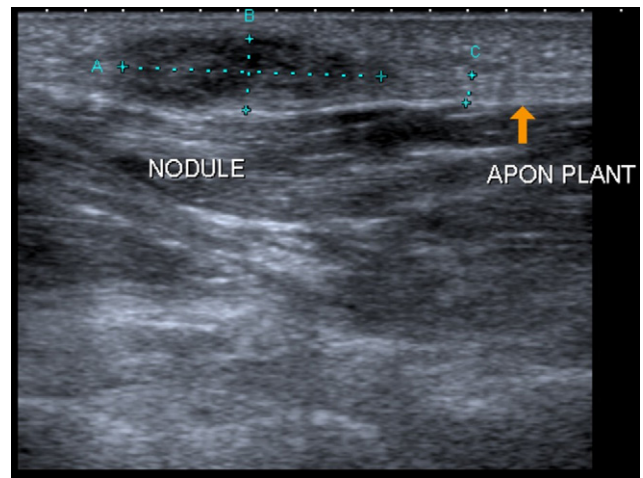


Figure 18. A 53-year-old woman who suffered from a painful nodule on the medial plantar surface of the foot. Sonography shows a nodule (between the dotted lines) attached to the plantar aponeurosis (orange arrow): plantar fibromatosis or Ledderhose’s disease.

thought to ideally require surgical removal although some advise the use of local steroid injections. Radiotherapy has sometimes been used postoperatively but functional sequelae are possible.

Deep fibromatoses

Desmoid tumours

These originate in the connective tissue of muscle, fascias, or aponeuroses, and they mainly affect young adults. There are three possible types: intra-abdominal, abdominal wall, or extra-abdominal. These types all have the same sonographic features: they are often quite large by the time they are investigated, with irregular finely spiculated margins, with thin peripheral extensions (Fig. 19). Their echostructure is solid, heterogeneous, and is made up of alternate linear zones or strips, that are hyper and hypoechoic, and of variable relative proportions. This seems to

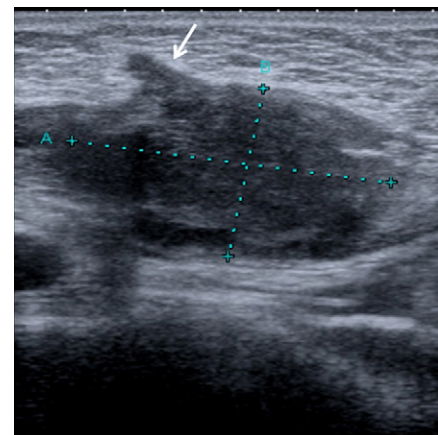


Figure 19. Desmoid tumour of the abdominal wall (rectus abdominis muscle) in a young 26-year-old woman. Sonogram in the axial plane demonstrates a wedge-shaped spiculated extension radiating between the intermuscular fascia (white arrow): very useful diagnostic sign.

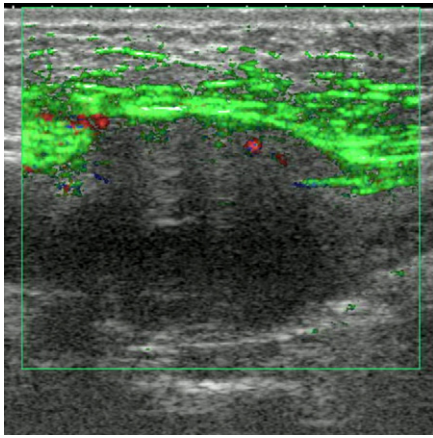


Figure 20. After administration of a contrast agent, minimal enhancement of the anterior part of the mass: a finding usually reported in inactive desmoid tumours. This particular lesion is situated in the rectus abdominus muscle of the abdominal wall (Same case as in Fig. 22).

correlate to the composition of the tissue, which is made up of cells (echogenic), fibres, and collagen (hypoechoic). On Doppler studies, vascularisation made up of a number of diffuse components is nearly always present. Findings vary on contrast-enhanced sonography. Although they can be considered to always enhance, this enhancement may be minimal (Fig. 20) or it can on the contrary be very intense (Fig. 21). This difference is doubtless related to the progression of the tumour. (We have observed that significant contrast uptake is seen in tumours that are often clinically tender and that are growing in size in spite of treatment). Certainly, these findings are similar to those of the highly aggressive lesions, a group to which desmoid tumours do not belong (they do have a tendency to relapse locally but they do not cause metastases). Both clinical (predisposition) and sonographic factors (spiculated margins, fine peripheral extensions, echostructure made up of alternate zones of

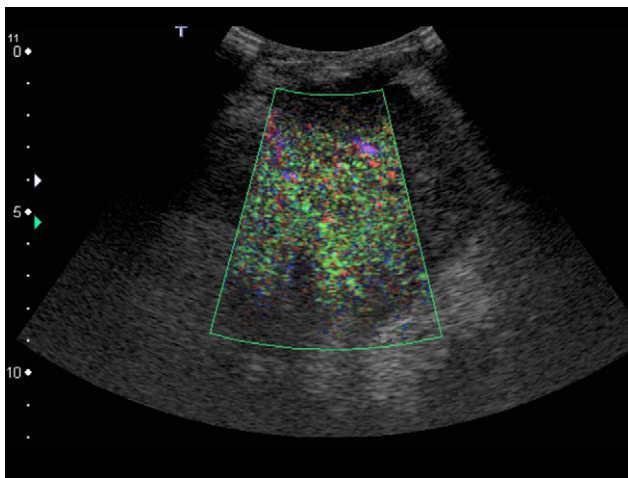


Figure 21. Quick and considerable contrast uptake in a desmoid tumour of the shoulder that had been causing the patient significant pain for around one month. The patient also reported a sensation of increased volume. This is a finding reported in “flare-ups” of desmoid tumours.

high and low echogenicity) can all be arguments in favour of a relatively reassuring diagnosis. Nonetheless, this does not mean that MRI and biopsy are not required for diagnostic confirmation [19,22,23].

Intra-abdominal fibromatoses

These are not tumours of the musculoskeletal system in the strictest sense. They are usually isolated, but can sometimes (in 15% of cases) be associated with familial adenomatous polyposis (Gardner’s syndrome). In these patients, localisation to the muscles or aponeuroses can also be seen [24].

Abdominal fibromatosis

This lesion presents a distinctive set of findings. It affects women and is very often connected to childbearing, occurring during or very soon after a pregnancy (within a year after the birth). It is known to be hormone-dependent since oestrogens seem to promote the proliferation of fibroblasts. The internal oblique and rectus abdominus muscles are the ones most commonly affected [25].

Extra-abdominal fibromatoses

These lesions originate close to the fascias of the shoulder muscles, chest wall, back, thighs, and in the knee area [24]. They produce the same sonography findings as are usually seen in desmoid tumours. They may be single or multiple (in over 15% of cases), and the younger the patient is, the more like they are to relapse [23].

Fibrosarcomas

A fibrosarcoma is a malignant proliferation of fibroblasts that are predominantly sited on the trunk and the extremities. They have the non-specific sonographic appearance of a highly aggressive lesion [6,14]. These lesions enhance after contrast injection. Histology and, to a lesser extent, MRI are used to distinguish them from other malignant lesions and even from some desmoid tumours, which can produce similar findings on sonography.

Fibrohistiocytic tumours and pseudotumours

Giant cell tumours of tendon sheath

This is a nodular form of pigmented villonodular synovitis [14]. It is firmly fixed to the underlying tendon sheath (Fig. 22). Usually found on the hand (it is one of the most common soft tissue lesions in this site), the wrist, or the volar aspect of the fingers close to an interphalangeal joint, it is seen in adults between their third and fifth decade, with a slight predominance in females. It is less commonly found in the knee or ankle. It is fusiform, well-defined, more echogenic than muscle, homogenous, and shows no cystic areas or calcification. It is not interdependent from the tendon fibres and when the fingers are moved, this can be nicely demonstrated during a dynamic examination. Sonography is the ideal modality for this purpose [2,4,13,26]. Doppler sonography can in rare cases show slight vascularisation. These lesions are non-enhancing.

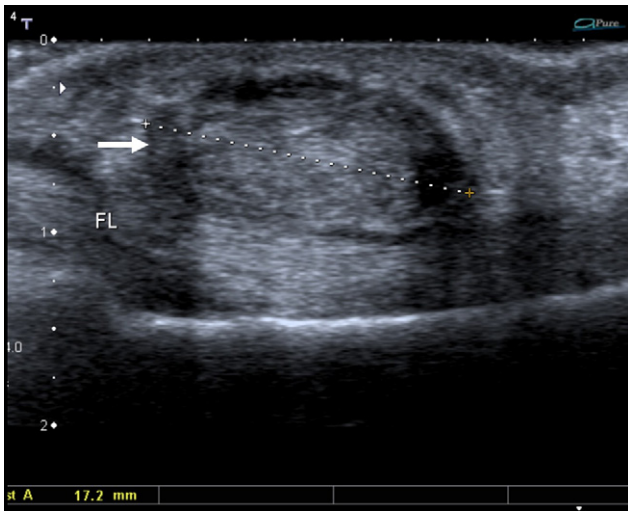


Figure 22. A 50-year-old male. Painless and non-progressive tumefaction of the anterior aspect of a finger. Sonography showed a solid, ovoid, well-defined, avascular formation (white arrow) ‘sitting’ on the tendon sheath of the superficial flexor of the finger (FL): giant cell tumour of the tendon sheath.

Benign fibrous histiocytoma

This is a hypoechoic, heterogeneous lesion with slight vascularisation. It is impossible to distinguish from a low-grade malignant lesion. MRI and biopsy are essential [27].

Malignant fibrous histiocytoma

This lesion used to be considered to be the most common malignant tumour in adults until the WHO recategorised it as a high-grade pleomorphic sarcoma (with the sub-groups: fibrous tissue and histiocyte predominant, giant cell predominant and inflammatory predominant) [4]. It has the sonographic features of an aggressive tumour, which means a biopsy is required as soon as possible.

A specific case of a benign pseudotumour: Xanthoma

A xanthoma is a localised proliferation of lipid-laden histiocytes that develops in patients with hypercholesterolaemia. They are usually known of, can be single or multiple, and are found in the cutaneous or subcutaneous layers, synovium, tendons, and in very rare cases, the bone [5,14]. It is usually caused by a reactive process rather than being a true neoplasm. On sonography, the classic findings are a tendon that appears ‘black and white’ and nodular on axial views, with a fibrillar pattern on the long axis [2]. Unusual features can be found (Fig. 23): solid formation, hypoechoic with lobulated margins, located on a tendon and sometimes causing bone remodeling, and avascular, both before and after administration of a contrast agent. In these atypical forms, laboratory tests for background information and, where necessary, a biopsy will shed light on these findings.

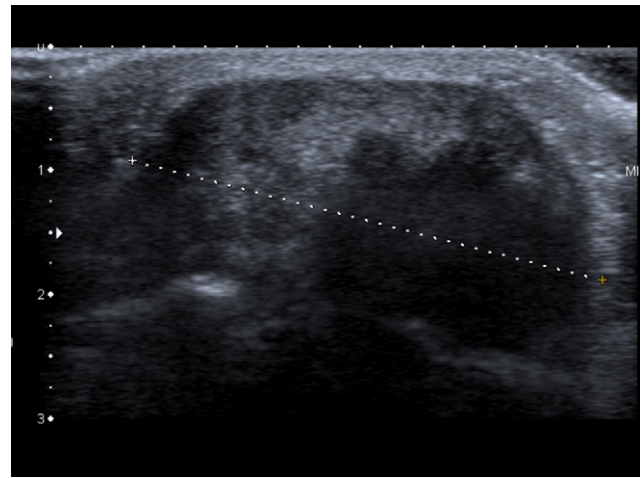


Figure 23. Sonography: axial view of the tibial insertion of the patellar tendon. Large, solid formation that is hypoechoic, painless, non-progressive, not vascularised either on Doppler imaging or after contrast agent administration, that ‘blends’ with the patellar tendon, in a 50-year-old female with hypercholesterolaemia. Biopsy confirmed a xanthoma.

Tumours and pseudotumours of the peripheral nerves

Schwannoma

These develop in the neck (vagus nerve, sympathetic nervous system) and the limbs (nerves of the flexor muscles) originating from the cells of the nerve sheath (meaning they have an eccentric growth with respect to the central nerve axis). Adults between the ages of twenty and fifty are affected, with equal distribution in both sexes [28,29].

Neurofibroma

Neurofibromas arise from constituents within the nerve (so they are centred on the nerve). They are usually single and affect young men and women in equal numbers, between the ages of twenty and thirty. In some rare cases (around 10%), they are associated with neurofibromatosis type 1.

Sonographic features are almost identical: fusiform, solid, moderately echogenic, sometimes with small, well-defined cystic areas, and an inconsistent finding is posterior enhancement. The essential point for diagnosis is a connection between the lesion and a nerve, which should ideally be demonstrated in real time (Fig. 24). Whether the lesion is eccentric to the nerve (schwannoma) or not (neurofibroma) seems to us to be a merely theoretical distinction in practical sonography. They are both generally avascular on Doppler imaging, and there is no contrast enhancement (in a few cases, it is possible to find minimal vascularisation on Doppler imaging, just as contrast enhancement can also be seen in rare cases) [29].

Morton neuroma

This term is misleading because this is in fact fibrosis of an intermetatarsal plantar nerve. It is mechanical in origin,

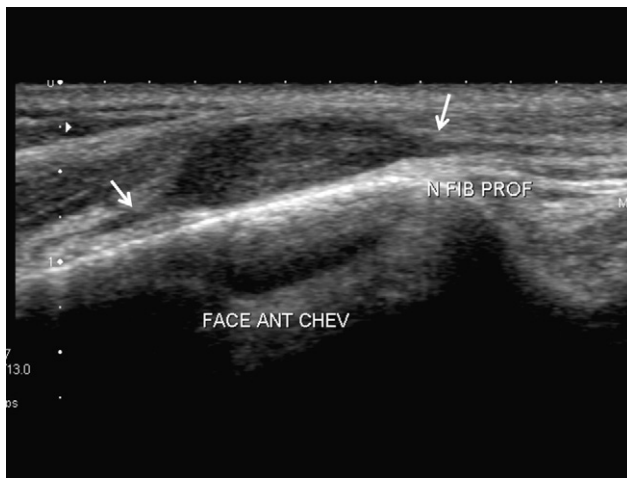


Figure 24. Longitudinal view of the anterior surface of the ankle in a 20-year-old woman who reported a tender nodule in this area, which was especially noticeable on mobilisation. Sonography did indeed demonstrate this small fusiform mass, which was solid, slightly echogenic, and without hypervascularisation shown either on Doppler imaging or after contrast agent administration (images not shown). The essential sign is present, in that it reattaches to the nerve at both extremities (white arrows): most likely a schwannoma.

being secondary to repeated micro trauma. There is a clear predominance in females. It causes pain on weight-bearing that is relieved by rest. It usually cannot be felt on palpation. Single, multiple, and bilateral forms are all found. It is investigated using the dorsal approach (with the help of Mulder's sign, which consists of firmly compressing the plantar surface of the forefoot while exerting a lateral pressure on the metatarsal heads) or the plantar approach, (the opposite finger is used to compress the dorsal side of the intermetatarsal space opposite to the probe). It is nearly always sited in the second or third intermetatarsal space. It is a bulbous and roughly oval formation that is hypoechoic, with indistinct margins, seen on longitudinal views to be positioned between the two nerves that are involved [30]. There is no vascularisation seen either before or after contrast agent administration (Fig. 25). It can be associated with intermetatarsal-phalangeal bursitis and this must not be allowed to mask a Morton neuroma [31].

Post-traumatic scar neuromas secondary to resection of the nerve must be mentioned, and these can be considered together with forms seen after amputation.

Malignant tumours of the peripheral nerve sheaths

These tumours are often, but not always (25–70% of cases), associated with neurofibromatosis, and they usually affect the major nerve networks (sciatic nerve, brachial plexus, or sacral plexus) [14]. They affect men and women in equal measure, except for those connected to neurofibromatosis type 1 (NF 1) in which there is a clear predominance in males. Usually developing between ages twenty and fifty, it is often seen earlier in these patients. These lesions are mainly deteriorating neurofibromas and a transformation

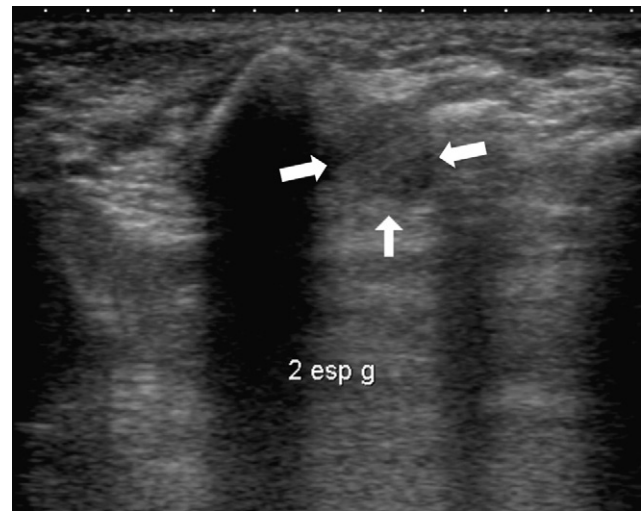


Figure 25. A 58-year-old female, complaining of diffuse forefoot pain on weight-bearing for more than one year, and difficulty putting on her shoes. Axial sonogram views of the area, taken both from the dorsal and plantar approaches, show a small, solid, hypoechoic formation (white arrow) of the second intermetatarsal space, that was linked to a plantar digital nerve, with no vascularisation either on Doppler imaging or with contrast-enhancement (images not shown).

into schwannoma is rare. They are often a source of pain along the nerve path (sciatica), and on sonography they appear as a large mass (with a diameter of several centimetres) that is solid, well-defined, heterogeneous, with several areas of necrosis, that is also continuous with the nerve structure it originates from [18]. Doppler imaging usually shows vascularisation within the lesion (except when there is significant necrosis) and, most importantly, use of a contrast agent demonstrates a warning sign as it shows intense and early enhancement, meaning that a biopsy is indicated (Fig. 26).

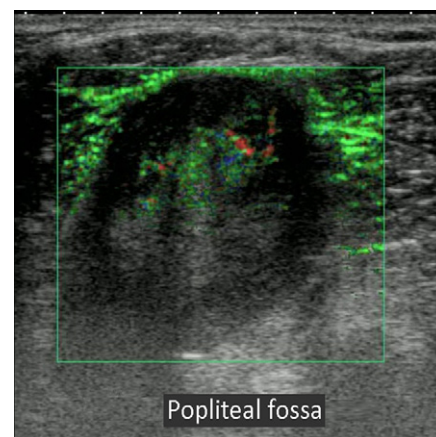


Figure 26. Large, painful and fast progressing tumour of the popliteal fossa, connected to the tibial nerve in a 20-year-old male with neurofibromatosis type 1. The transverse sonogram view of this region showed a discrete tumour with contrast uptake (aggressive lesion). This is a schwannosarcoma.

Extraskkeletal osseous and cartilaginous tumours

Benign tumours or pseudotumours

Myositis ossificans circumscripta

This is a benign heterotopic ossification of the soft tissue that is sometimes due to trauma. It is made up of fibroblasts and myofibroblasts, which explains why it is currently classified under fibroblastic/myofibroblastic tumours (we preferred to consider it in this category because this seems to us to correspond to the reality of its sonographic features) [4]. Osteoblasts and chondrocytes subsequently appear and ultimately, mature bone. The clinical picture sometimes consists of pain and local inflammation, or the lesion may be inactive, and discovered accidentally. Although in the early stages misdiagnosis may be made on sonography (it is thought that, if carried out an early stage, sonography sometimes shows a small hypoechoic zone within the muscle that is surrounded by vessels, findings that could also be seen in an early, aggressive extraskkeletal tumour), when it is carried out later, it is by contrast able to show, with good sensitivity, crescent-shaped peripheral calcifications that generate a shadow cone and prevent exhaustive investigation of the lesion (Fig. 27a) [6]. It may be situated in the subcutaneous tissue (panniculitis ossificans) or in the fascias (fasciitis ossificans) and of course, in the skeletal muscle, which is the reason for the name it bears myositis ossificans. When local inflammation is present, this explains the hypervascularisation seen on color Doppler sonography, as well as the rapid and intense contrast uptake (Fig. 27b). When it becomes asymptomatic there is only moderate vascularisation at the point of contact with the calcifications. A CT scan is the best modality for examining the lesion in its entirety.

There are other lesions that arise from periosteum such as Nora's tumour (bizarre parosteal osteochondromatous proliferation), which can be uncovered on sonography, but accurate diagnosis of these does rely on other techniques [32].

Chondroma and osteochondroma of the soft tissue

These are small cartilaginous nodules that sometimes also ossified and they are predominantly found in the hands and feet. They are more usually investigated with standard radiography and CT [5,14]. If there is exostosis, sonography may prove to be useful to measure the thickness of the cartilaginous layer, as this can give an idea of its aggressiveness [2].

Malignant tumours

Extraskkeletal chondrosarcomas and osteosarcomas: these are rare and CT and MRI are the most suitable investigations [5,14].

Vascular and lymphatic system tumours

This category includes haemangiomas and lymphangiomas, whether benign or intermediately malignant, haemangi endotheliomas and haemangiopericytomas, and the malignant processes angiosarcoma and Kaposi's sarcoma. We also discuss a specific lesion in this section: the glomus tumour.

Benign tumours

Haemangiomas [14,33,34]

Haemangiomas are the most common hamartomas of the soft tissue. They make up 7% of benign tumours. They are usually identified before the age of three and are the lesion most often found in infants and children. They can also be seen in adolescents and adults. They predominantly affect females. Their location can be superficial or deep, and in the latter case they are nearly always intramuscular. On histology, they are made up of a wide range of tissue types. Anatomical pathology classes them according to the dominant vessel type: capillary, cavernous, arteriovenous, or venous. A capillary haemangioma is superficial, situated in the skin and subcutaneous tissue, and is reported in infants

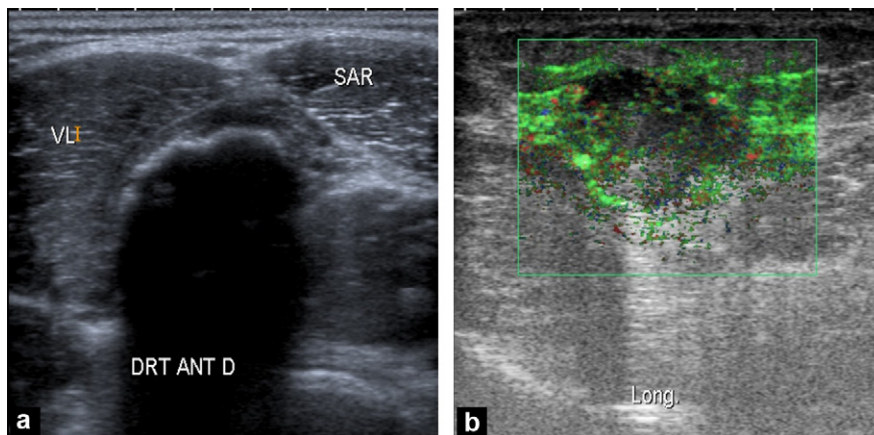


Figure 27. a: transverse sonogram view of the proximal thigh in a 40-year-old male who reported pain. Crescent-shaped intramuscular calcification producing a shadow cone and preventing overall visualisation of the lesion: myositis ossificans; b: early and considerable contrast uptake in a young woman presenting a picture of soft tissue inflammation of the internal aspect of the thigh: flare-up of myositis ossificans related to progression demonstrated by anatomical pathology explorations.

and young children. It involutes before the age of seven and there is no need for imaging.

A cavernous haemangioma is intramuscular, made up of dilated blood-filled spaces that are lined by a flattened endothelium. They are mainly seen in young children and adults. They do not involute. This type of haemangioma is the one most often explored using imaging. Sonography shows a formation bordered by an echogenic, thin, and slightly irregular boundary (fatty in nature) containing a complex echostructure, that is non-homogenous, being made up of hyperechoic (lipids) or isoechoic areas (smooth muscle, fibrosis, hemosiderin) amongst which are scattered zones of fluid and vasculature, which are demonstrated well by all types of Doppler imaging. Spectral tracings of systolic-diastolic flow that are indicative of arteriovenous shunts are a classic finding. Phleboliths and dystrophic calcifications of an organised thrombus may also be identified (Fig. 28a and b). These lesions are sometimes very large and spread into the adjacent soft tissue, or even the bone. They are contrast-enhancing, though this is seen relatively late (around 1 minute 30 s) (Fig. 28c).

An arteriovenous haemangioma reproduces the model of the foetal capillary network. They are sited in the soft tissue in young children. Characterised by increased blood output, they can sometimes cause malformations (enlargement of the extremities, venous distension etc.).

A venous haemangioma is made up of vessels with thick muscular walls. They are usually deep (retroperitoneum, mesentery), meaning they are almost never reported in the musculoskeletal system.

Lymphangiomas [14,35]

Lymphangiomas are made up of "excluded" lymphoid tissue that does not communicate with the lymphatic system and they are lined by lymphatic endothelium. They are classified according to the size of the vessel: capillary, cavernous, and, the most common type, cystic hygroma. The latter are usually present from birth and are always diagnosed before the age of two. They are usually found in the axilla, sub-mandibular region, and posterior neck (from where they can spread to the mediastinum).

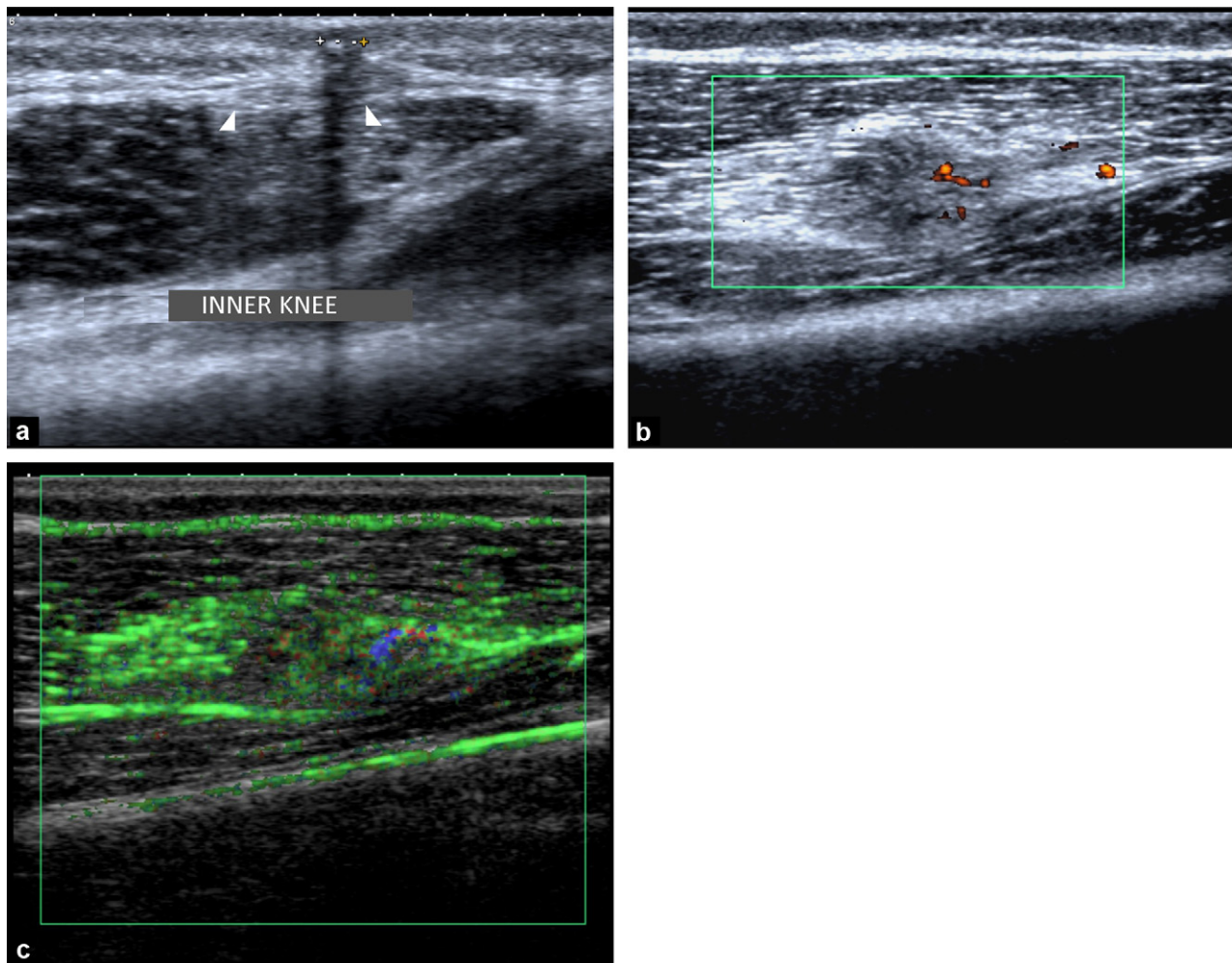


Figure 28. a: young 14-year-old male who reported asymmetrically sized thighs and pain on mobilisation. Sonography showed a deep, intramuscular formation that was immediately sub-aponeurotic (arrowheads) with a phlebolith (measurement shown by small crosses): intramuscular haemangioma. Hypervascularisation shown on color Doppler (b) and late contrast uptake (around 1 min 30 s) (c) after injection.

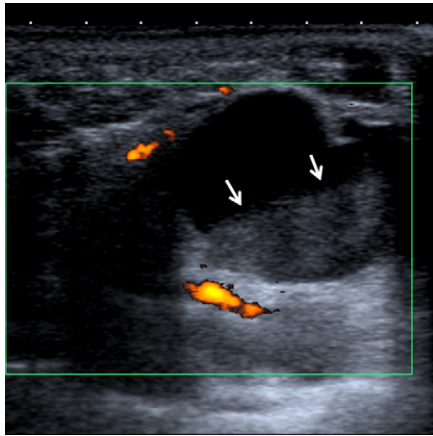


Figure 29. Haemolympangioma of the sub-mandibular triangle, already known of, in a 14-year-old girl who was examined due to acute and painful swelling. Visualisation of the lesion showed an air-fluid level (white arrows) and echogenic sediment: very likely to be intralesional bleeding.

Sonography demonstrates a cystic mass with multiple septations, the septa being of variable thickness. A few solid components or septations may be seen. Complications can develop: infections, causing the contents to appear more echogenic, bleeding, in which an air-fluid level may appear, or even in rare cases, rupture (Fig. 29).

Intermediate malignancy tumours

A haemangiopericytoma is a proliferation of endothelial vascular cells that varies in terms of site and topography (deep or superficial) [2,14]. A haemangiopericytoma is made up of pericytes, contractile cells that surround the capillaries and post-capillary venules, and it affects middle-aged adults with no predominance in either sex, developing mainly on the extremities of the lower limbs and the retroperitoneum [14]. Features on sonography are non-specific.

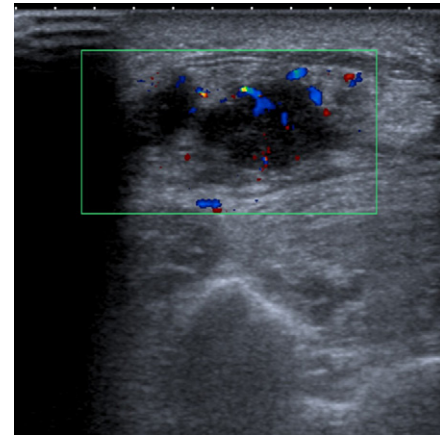


Figure 30. Fast progressing mass of the posteroinferior part of the leg in an 80-year-old male. Sonography showed a lesion with irregular margins, areas of necrosis, and anarchic hypervascularisation: this lesion presented significant uptake of sonographic contrast material (image not shown): aggressive lesion: biopsy confirmed leiomyosarcoma.

Malignant tumours

Angiosarcomas, whether superficial or deep, affect older patients and are twice as common in men. Chronic lymphoedema is a predisposing factor [14,35].

Kaposi’s sarcoma is a malignant cutaneous vascular proliferation associated with a viral infection. There are four different clinical contexts: chronic, lymphadenopathic, in transplant patients, and in patients with AIDS, for whom the prognosis is poorer.

On sonography, it is impossible to determine whether any of this group of tumours is of intermediate or certain malignancy because there are no specific signs. MRI has greater possibilities. Biopsy is essential [14].

Glomus tumours

This is a benign neoplasm that arises from a neuromyoarterial glomus body. It is usually found on the dorsal surface of

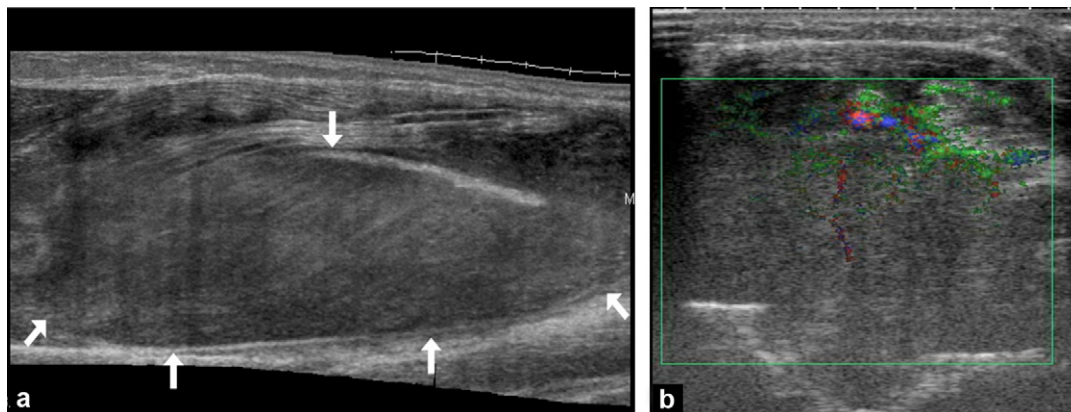


Figure 31. Painful, fast growing mass of the anterior surface of the arm in a 14-year-old male: a: sonography demonstrated a discrete, solid mass with a feathered appearance occupying the anterior brachial compartment; b: after administration of contrast agent, the tumour quickly showed enhancement: aggressive lesion of the anterior compartment of the arm in an adolescent: rhabdomyosarcoma confirmed on biopsy.



Figure 32. Sonography in a 65-year-old female presenting a large tumefaction on the external surface of the right thigh: ovoid, intramuscular formation with an almost-fluid echostructure and no contrast-enhancement seen. Histology confirmed a myxoma.

the fingers, under the nails. Other possible locations are the wrist, forearm, and foot. There is a clear predominance in females. Clinically, there is a small bluish or reddish nodule that presents paroxysmal pain or pain on pressure and they are often sensitive to cold. It is difficult to make a diagnosis using sonography when the tumour is small as the nail can act as a barrier to ultrasound waves. When sonography does find this tumour, it is in the form of a small sub-ungual hypoechoic mass that shows hypervascularisation on Doppler imaging [5,14].

Tumours and pseudotumours of the muscles

Smooth muscle

Benign tumours: leiomyoma, usually small, superficial, cutaneous or subcutaneous, not investigated by imaging.

Malignant tumours: leiomyosarcomas, which have no specific characteristics and the only feature to look for, especially if it is a venous lesion, is a point of contact between the tumour and vascular wall that the process could have originated from [2,14]. The usual features are once again those seen in aggressive lesions (Fig. 30) [2].

Striated muscle

Benign tumours: rhabdomyoma is very rare and affects middle-aged adults with a clear male predominance [2,5,14]. It is usually sited in the head or neck and has no particular features on sonography.

Malignant tumours: rhabdomyosarcoma, which is the most common soft tissue tumour in children [2,5,14]. Its features on sonography are those seen in all aggressive tumours (Fig. 31a and b). MRI and ultimately biopsy are essential.

We include under this section muscular abnormalities that are in fact anatomical variants: supernumerary muscles that take on the clinical appearance of a tumefaction, the true cause of which will generally be quickly identified on sonography [36].

Tumours of uncertain differentiation

Myxoma

This is a benign mesenchymal tumour seen in adults that is rich in stroma, avascular, myxoid, and has a minimal cellular component. It is predominantly sited in the muscles of the thighs, shoulders, buttocks, and arms [37].

On sonography, it appears as a well-defined ovoid mass within the muscle that is highly hypoechoic, almost fluid, not vascularised on Doppler imaging (Fig. 32), and is non-enhancing. There is an association with polyostotic fibrous dysplasia, a presentation that constitutes Mazabraud's syndrome.

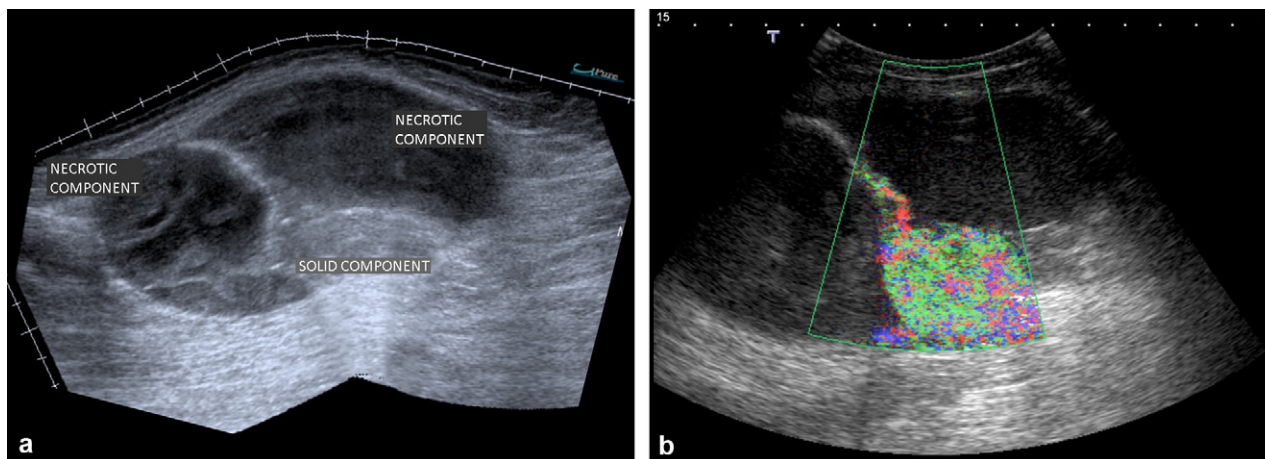


Figure 33. Fast progressing “mass” of the internal surface of the thigh in a 55-year-old female. Sonography shows a large, non-homogenous formation that is made up of both solid and fluid zones, separated by thick septations (a). After contrast agent administration, there is fast and significant contrast uptake in the solid section and the septations (b). Synovial sarcoma identified on histology investigations.

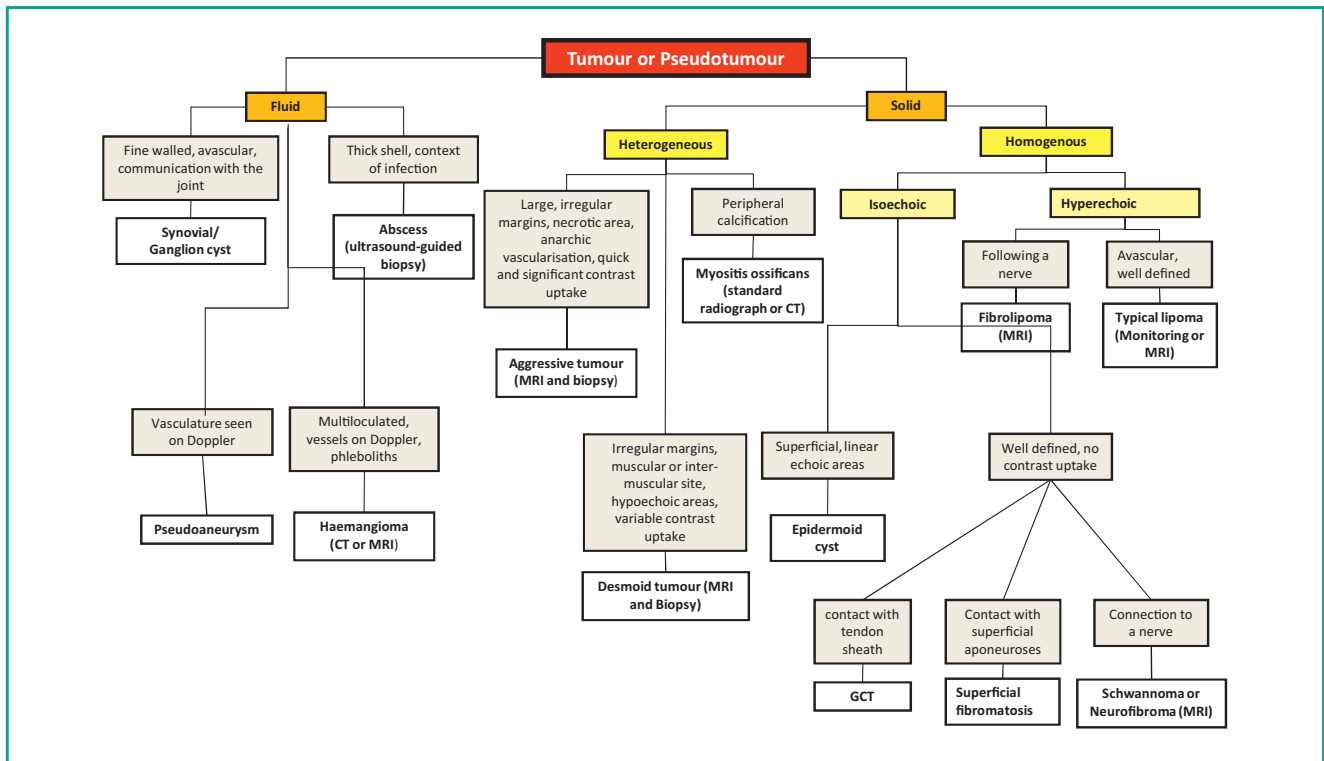


Figure 34. Flow-chart.

Synovial sarcoma

The name synovial sarcoma is misleading. This lesion originates in the para-articular tissue and is predominantly found in the lower limbs. It is fusiform or lobulated, hypoechoic, and shows anarchic hypervascularisation; in fact it has the sonographic features of an aggressive lesion, without being specific (Fig. 33a and b) [6, 14, 38]. MRI and especially biopsy will once again provide the solution for the diagnosis. Sonography is useful and can be used to investigate recurrence, when it shows a small nodular, hypoechoic mass under the scar tissue.

Metastases

Metastases in the soft tissue are uncommon, and they originate from the lungs, kidneys, or gastrointestinal system. Knowledge of a primary malignancy will assist with diagnosis. They produce a picture of a discrete tissue mass that varies in size and demonstrates hypervascularisation on Doppler imaging. These lesions do enhance. If no primary lesion is known of, a PET Scan could prove to be very useful [6].

Conclusion

Currently sonography occupies an important place in the investigation of soft tissue tumours. It should initiate the imaging investigations. Taken together with the clinical findings (patient’s age, progression and topography of the lesion,

context of infection or bleeding) it can, in quite a number of cases (simple lipoma, synovial cyst, vascular malformation, benign tumour of peripheral nerve sheaths, elastofibroma dorsi, palmar or plantar fibromatosis, haematoma, abscess), provide enough information for a conclusive diagnosis, as long as close monitoring is scheduled. In other cases, taking the clinical picture and lesion topography into consideration, it may lead to a probable diagnosis: giant cell tumour of the tendon sheath, desmoid tumour, myositis ossificans. Further MRI investigation, or biopsy are sometimes necessary, to confirm the diagnosis.

In still other cases, sonography can provide findings that are suggestive of an aggressive process, or reveal the presence of atypical features (anarchic vascularisation). It is then down to other investigations (always MRI, sometimes CT) to find further features of diagnostic value (Fig. 34).

It remains an open question what the contribution of innovations in sonographic technology will be (3D, elastography, contrast), further investigation is needed to ascertain the diagnostic benefits of these techniques.

Later, there is nearly always a return to sonography to guide with relative ease a needle biopsy of these generally accessible lesions, as this allows a histological diagnosis to be made and a suitable treatment plan to be set out.

Finally, sonography is useful after treatment for monitoring tumoral lesions, by assessing and checking on the response to treatment, based on inactivity at the site as well as under and around the scar tissue, although it is important to be aware that a lesion measuring less than one centimetre can escape vigilant observation due to postoperative fibrosis.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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