Imaging of Benign Lipomatous Tumours of The Limbs In Children

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Imagerie des tumeurs graisseuses bénignes des membres chez l'enfant

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RÉSUMÉ

Prérequis : Les tumeurs graisseuses des membres chez l'enfant sont rares et sont souvent bénignes. Le lipoblastome est la tumeur la plus commune.
But : Le but de ce travail est d'illustrer les aspects cliniques, radiologiques et histologiques des principales tumeurs graisseuses des membres chez l'enfant.
Méthodes: Revue de la littérature.
Résultats : Etude descriptive de cas pédiatriques de tumeurs graisseuses bénignes des membres chez l'enfant.
Conclusion : L'imagerie peut fournir des éléments essentiels pour le diagnostic.
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SUMMARY

Background : Lipomatous tumours of the limbs in children are rare, and lipoblastoma is the most common soft tissue tumour. Most of them have typical imaging features, but their clinical presentation and their management may vary, depending on the exact histological subtype.
The aim of our study is to illustrate the main clinical, radiological and histological features of the different benign lipomatous tumours in children.
Methods: review of the literature.
Results: it is about a descriptive study of paediatric cases of benign lipomatous tumours of limb in children.
Conclusion: The imaging findings are helpful and can provide essential components for the diagnosis.

Mots-clés
Tumeurs graisseuses, membres, enfant, imagerie.

Key-words
Lipomatous tumour, limbs, children, imaging.

Lipomatous tumours of the limbs in children are rare and generally develop within the soft tissues. They are more often encountered during the first years of life and they are more frequent in boys than in girls [1, 2]. Lipomatous tumours involving soft tissue are generally benign, especially before 5 years of age; their classification is based on their morphology and cell type [3, 4, 5].

Lipoblastomatous lesions occur almost exclusively in children and contain a spectrum of fat cells ranging from immature lipoblasts to mature adipocytes. Lipomas and lipomatosis consist of mature adipocytes [1, 2].

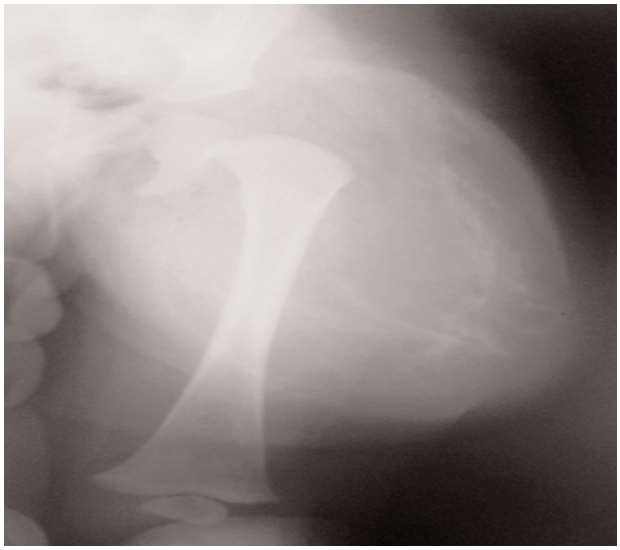
Their fatty contents is easily identified with various imaging modalities, which can in some cases give an indication on their histological nature, and in all cases make it possible to specify their extension and to direct the treatment strategy [6].

COMMON CHARACTERISTICS OF A LIPOMATOUS SOFT TISSUE TUMOUR OF THE LIMBS:

The most frequent clinical sign is the observation of a mass of variable consistency, limits and volume, which may be painless or moderately painful [7]. Symptoms are different depending upon the size and location of the tumour. The imaging strategy includes as the first line the combination of plain radiography and color Doppler sonography; this can be complemented by MRI if necessary. It assists in guiding appropriate management: conservative versus surgery. CT entails significant radiation, and it no longer used routinely.

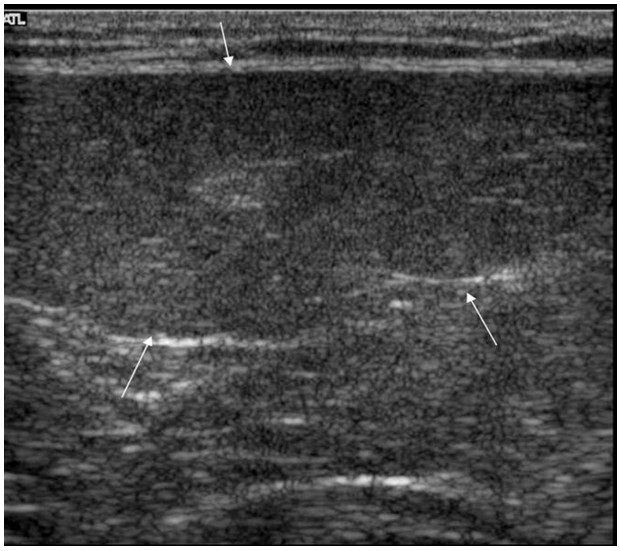
Plain radiographs show a radiolucent soft tissue mass with radiodensity similar to subcutaneous fat (Fig. 1) with sometimes cortical thickening of an underlying bone, exceptionally bone erosion [8, 9].

Figure 1 : Plain radiograph showing a large radiolucent soft- tissue tumour in a 7-month-old girl, in the posterior left thigh.



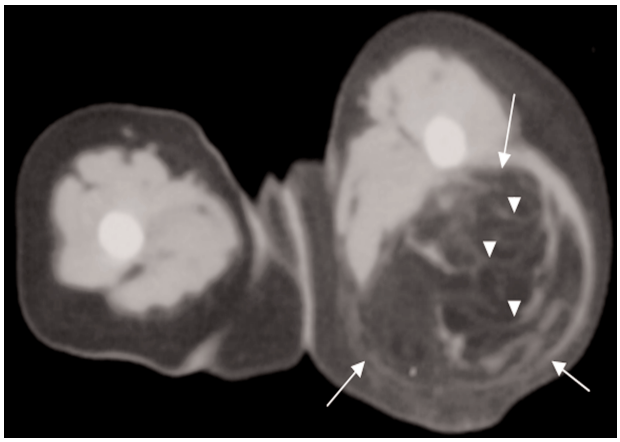
On sonography, a fatty tumour appears as oval, mostly well-defined, homogeneous masses with iso-, hypo, or hyperechoic presentation without detectable blood flow in color Doppler (Fig. 2).

Figure 2 : Longitudinal sonographic image shows an elongated, well delineated and homogeneous hypoechoic mass (arrows).



On CT, it is a hypodense mass with negative densities (-60 to -120 UH), sometimes septated and surrounded by a pseudo capsule; the septa and pseudocapsule show slightly enhanced density after contrast injection (Fig. 3).

Figure 3 : Rapidly growing deep mass tumour in a 7-month-old girl: Non contrast axial CT image shows a lobulated large mass in the posterior left thigh with fat density and well-defined margins (arrows). Thin septations within the mass are noted (arrowheads).



MRI provides the most reliable noninvasive assessment of soft tissue masses. Fatty tumours had high signal intensity on MR T1-weighted images and relative decreasing signal on T2-weighted images with signal loss on fat saturation pulse sequences (Fig 4). Sometimes they may contain thin septations of increased T2 signal which have moderate if any enhancement [2, 10]. The final diagnosis is done by pathology and cytology (fig 5). Histologically, fatty tumours consist of lipomatous cells surrounded by a pseudo capsule. Surgical excision is the treatment of choice and the prognosis is excellent.

Figure 4 : Intramuscular fatty tumour developed in the right leg in a 3-year-old boy. a) Axial MR T1-weighted image shows a homogenous high signal intensity mass (arrows), isointense to subcutaneous fat with thin fibrous septas and a surrounding capsule of low signal intensity. b) Axial MR T2-weighted image shows an intermediate signal mass. c) Axial fat-suppressed spin-echo T1-weighted image confirms the fatty nature of the tumour with complete absence of signal.

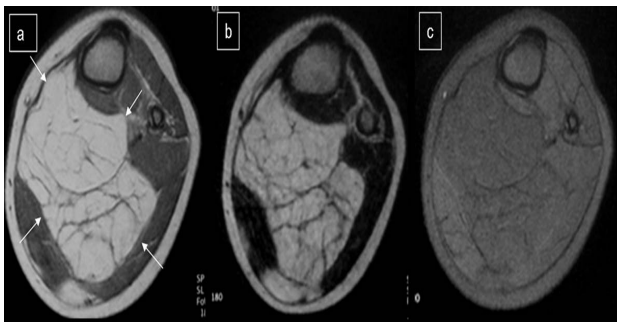
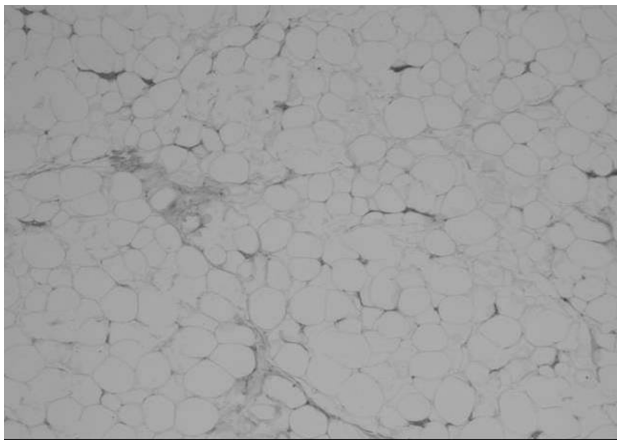


Figure 5 : Pathological study of typical lipoma demonstrates napes of adipocytes with thin conjonctivo-vascular septa.



CHARACTERISTICS OF THE PRINCIPAL INFANTILE TUMOURS OF SOFT TISSUES OF THE LIMBS:

Lipoblastoma: is a rare neoplasm of embryonal fat cells. Two clinical forms, with similar histological features, have been described. Lipoblastoma represents a well-encapsulated, isolated superficial (subcutaneous) lesion [9], whereas lipoblastomatosis refers to a more diffuse infiltrative process [11]. Some authors suggest the phrase "infiltrating lipoblastoma" for lesions with local invasion of muscles, analogous to "infiltrating lipoma" currently used to describe the mature infiltrating fatty lesion. The word "lipoblastomatosis" should be reserved for the more aggressive lesions that extend across anatomic planes and into different tissues [3].

The wide majority are detected in children under the age of 5 years, with a median age of 3 years [4]. Lipoblastomas usually present as rapidly growing masses most often located in the extremities [3, 12].

On MRI scans, they can have a more heterogeneous T1 appearance and may be comparatively hypointense to subcutaneous fat on T1-weight images which most likely correlates with the amount of lipoblastic tissue or fibrous tissue [1, 3, 13]. Lipoblasts have lower signal than lipocytes on T1-weight sequences [2]. This finding may be of potential value in differentiating lipoblastomatous from lipomatous lesions and from exceptional infantile liposarcoma [3, 4, 5, 14].

Histologically, lipoblastomas tend to appear as multiple lesions with a lobulated pattern due to connective tissue septae and embryonal lipoblasts at various stages of development in a myxoid stroma [4, 11].

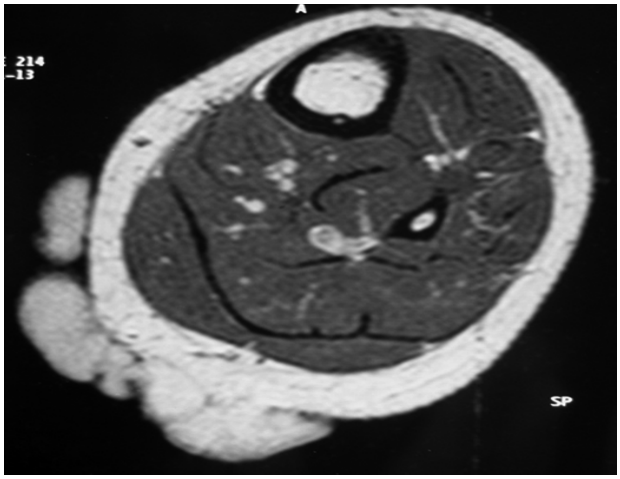
It is now well established that lipoblastoma is a benign tumour best treated by complete surgical excision and that a radical operation is not necessary and may be mutilating, especially in children [15]. A regular and periodic follow up is necessary for many years because the recurrence rates in local or marginal excision range from 12 % to 25 % in spite of complete excision [4, 11, 12, 15, 16].

These tumours have an excellent prognosis despite their potential to invade locally and to grow rapidly to a considerable size [4, 15].

Furthermore, the literature supports the lipoblastoma's capacity to mature from a histologic pattern composed of lipoblastes to a pattern composed solely of mature lipocytes [3, 17].

Simple lipoma: is a well-defined fatty tumour that usually arises in the subcutaneous tissue but can also have a deep-seated location in the subfascial compartment (inter muscular) or in other region such as the interdigital web (Fig. 6) [18].

Figure 6 : Axial MR T1-weighted images of the left leg in an 11-year-old boy showing a superficial lobulated mass, hyperintense continuing the sub cutaneous fat.



This tumour is more frequent in adults. Clinically, the diagnosis of a superficial lipoma is considered when confronted with a subcutaneous mass, more or less mollasse, of slow growth, moderate size and non painful.

Ultrasound finds all its interest in these surface localizations, it makes it possible to establish the diagnosis by showing a lengthened circumscribed mass, whose large axis is often parallel with that of the affected limb, measuring typically less than 5 cm, hyperechoic, non vascularised and depressing under

the pressure of the probe [19]. The surgical excision is done on the ultrasound data.

Ultasound interest is less important in deep-seated locations where the CT and better the MRI remains the examinations of choice and their findings closely resemble those sus described in " common characteristics of a lipomatous tumour of soft tissues of the limbs" [20].

The histological examination confirms the diagnosis by showing a well limited tumour, lobulated consisting in mature adipocytes with conjonctivo-vascular septas.

Infiltrating lipoma or infiltrating intramuscular lipomas: are so named because they infiltrate skeletal muscle. These tumours are extremely rare in the child population [3, 16, 21]. They tend to be circumscribed, but non encapsulated. In children, these lesions most often appear by a muscular distortion or enlargement of an entire extremity or a palpable mass [16, 21]. They are more easily found on contraction of the involved muscle [11]. Impingement or invasion of nerves can result in sensory or motor deficit [3]. The most common sites are large muscles of the extremities, especially those of the thigh, shoulder, and upper arm [11, 21, 22].

Plain radiographs show location in the muscular plane (Fig. 7a) with sometimes a feathery appearance caused by strands of skeletal muscle running through the mass [21].

Intramuscular lipomas tend to be uninodular on MRI scans (Fig. 7b) and although infiltrative they tend not to destroy the normal tissue around them [14]. Inhomogeneous signal relatively isointense with muscle may be noted within the lesion, probably reflecting muscle interspersed with fat and linear areas of hypo intensity with T1 and T2 weight correspond to fibrous tissue [3]. Intramuscular lipoma is made up by mature adipose tissue scantily laced with connective tissue and capillaries that extend beyond the muscle fascia into the intermuscular connective tissue spaces [11].

Complete surgical excision is recommended. Recurrence appears to be relating to an incomplete resection [11].

Other benign soft tissue lipomatous lesions

Hibernoma, chondroid lipoma, angiolipoma and myolipoma are subcutaneous or deep soft tissue masses occurring rarely in children. All of these have a heterogeneous appearance on imaging. Hibernoma is consisting of embryonic fat wich has signal intensity similar to that of fat on MRI with typical branching vascular structures [23].

On ultrasound, angiolipoma is echoic with ill defined outline and a vascularized component in color Doppler. Chondroid lipoma has a calcified component. These data are found in CT, MRI and on histologic evaluation.

Diffuse lipomatosis: is extremely rare and implies a more extensive process where infiltration by mature fatty tissue is not limited predominantly to muscular tissue but involves large portions of an extremity. An osseous hypertrophy is very often associated. The onset of lipomatosis is usually in the first 2 years of life. Plain films are exceedingly important to detect bone overgrowth which suggests the diagnosis, as is also the case in macrodystrophia lipomatosa and Proteus syndrome.

Flow chart while a hyperechoic mass

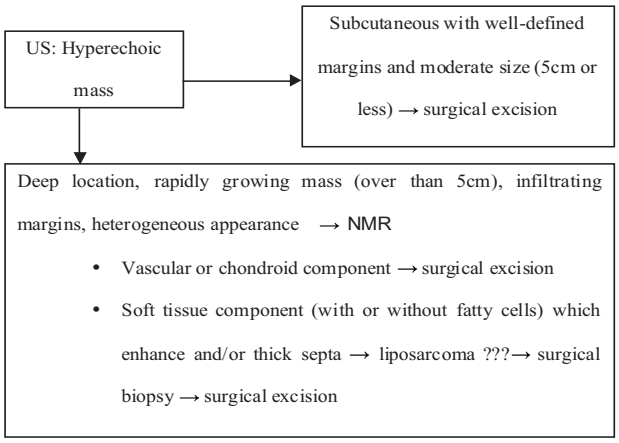
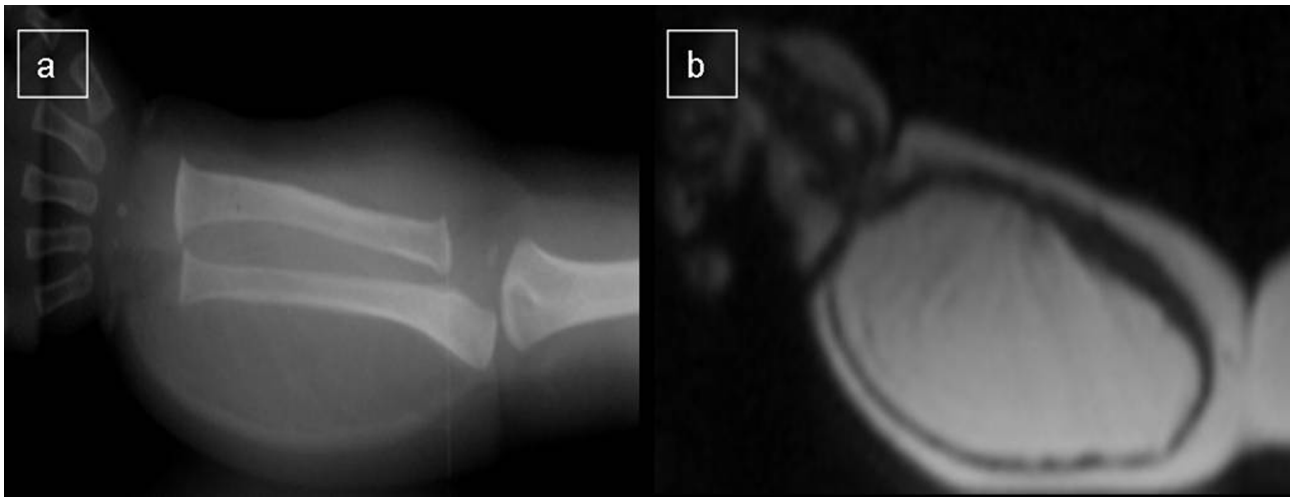


Figure 7 : Large deep mass of the right forearm in a 7-month-old boy. a) Standard radiograph showing a large soft tissue mass in the muscular plane pushing against the ulna. b) Coronal MR T1-weighted image showing the high signal intensity mass replacing the anterior-internal muscles of the right forearm.



DIFFERENTIAL DIAGNOSIS:

All the masses of the soft tissues which contain fat in children are in the differential diagnostic such as teratomas, deep hemangiomas after involution, lymphangioma, venous malformation, fibrous infantile hamartoma. Venous malformation may be characterized by blue mass, changing size with valsalva maneuver and phleboliths caused by stasis [24, 25].

Finally, the rare but difficult problem of well-differentiated liposarcoma which is exceedingly rare before 5 years of age, tend to be multinodular, more invasive and have a large amount of peripheral oedema [4, 5, 26, 14, 22, 27]. Furthermore, septations tend to be thicker and enhance to a greater extent, because often there are muscle fibers within these septa [28, 29]. Differentiation must be done on the basis of histologic evaluation.

CONCLUSION

Lipomatous tumours of the limbs in children are rare. Imaging findings are helpful and can provide essential components for the diagnosis, but their clinical presentation and their management may vary, depending on the exact histological subtype.

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