

## Imaging of lipoblastoma of the limbs in children

### Imagerie des lipoblastomes des membres chez l'enfant

Douira-Khoms W.<sup>1</sup>, Smida M.<sup>2</sup>, Sayed M.<sup>1</sup>, Louati H.<sup>1</sup>, Ben Hassine L.<sup>1</sup>, Lahmar L.<sup>1</sup>, Mrad K.<sup>3</sup>, Ben Ghachem M.<sup>2</sup>, Hammou A.<sup>4</sup>, Bellagha I.<sup>1</sup>

<sup>1</sup> Department of Pediatric Radiology. Tunis Children Hospital. 1007 Bab Saadoun - Tunis, Tunisia.

<sup>2</sup> Department of Pediatric Orthopedic Surgery. Tunis Children Hospital. 1007 Bab Saadoun - Tunis, Tunisia.

<sup>3</sup> Department of Pathology. Salah Azaiz Institute. Tunisia,

<sup>4</sup> National Center of Radiation Protection. 1007 Bab Saadoun - Tunis, Tunisia.

CORRESPONDENCE: **Dr Wièm DOUIRA-KHOMSI.**

Service de Radiologie Pédiatrique. Hôpital d'Enfants de Tunis. 1007 Place Bab Saadoun, Tunis - Tunisie

E-mail: khomsiwiem@yahoo.fr

#### ABSTRACT

Lipoblastoma is uncommon benign mesenchymal tumor arising from embryonic adipose tissue. The tumor occurs primarily in infancy and early childhood and commonly arises from the limbs. It has no malignant potential but may recur in cases of incomplete resection. Lipoblastoma may mimic other infantile tumors, including hemangioma, hibernoma, lipoma and liposarcoma, and correct diagnosis is necessary to ensure appropriate treatment. The imaging findings are helpful and can provide essential components for the diagnosis. In this paper, we present a review of the literature related to lipoblastoma of the limbs in children. We illustrate the main clinical, radiological and histological characteristics of this tumor.

#### RÉSUMÉ

Le lipoblastome est tumeur mésenchymateuse bénigne rare issue des cellules graisseuses embryonnaires. Cette tumeur touche souvent l'enfant de moins de 5 ans et se localise préférentiellement aux extrémités des membres. Elle ne présente aucun potentiel malin mais peut récidiver en cas d'exérèse insuffisante. Le lipoblastoma peut simuler d'autres tumeurs infantiles, y compris l'hémangiome, le hibernome, le lipome et le liposarcome, et le diagnostic de certitude est nécessaire pour assurer le traitement approprié. L'imagerie peut fournir des éléments essentiels pour le diagnostic. Dans ce travail, nous illustrons les principales caractéristiques cliniques, radiologiques et histologiques du lipoblastoma des membres chez les enfants avec revue de la littérature.

## I. INTRODUCTION

Lipoblastoma is a rare benign mesenchymal tumor arising from embryonic white adipocytes [1, 2]. Synonyms of this lesion are embryonic lipoma, fetal lipoma, lipoblastic tumor and congenital lipomatoid tumor [3]. Two forms of lipoblastoma have been described: a localized well-encapsulated mass (lipoblastoma) or a diffuse multicentric lesion (lipoblastomatosis) [4, 5]. The word "lipoblastomatosis" should be reserved for the more aggressive lesions that extend across anatomic planes and into different tissues [6].

## II. EPIDEMIOLOGY AND CLINICAL FEATURES

Lipoblastoma occurs almost exclusively in infancy and early childhood. It is typically encountered in infants less than 3 years of age; however, it has been reported in adults [1, 2, 7, 8]. The male-female ratio is 3/1 [1, 2]. The exact pathogenesis of this tumor has not been well established [9]. Lipoblastoma occurs mostly in the lower extremities but is also seen in the upper extremities and other body sites [2, 10, 11]. The circumscribed lipoblastoma is more common (approximately 70% of cases) and is located in the superficial soft tissues. The diffuse type lipoblastomatosis (about 30% of cases) has an infiltrative growth pattern that affects the subcutaneous tissue and underlying muscle [7]. Symptoms are different depending upon the size and location of the tumor. Lipoblastomas most commonly manifest as asymptomatic, painless, progressively growing masses in the superficial or subcutaneous soft tissue of the extremities [1, 10, 12]. A rapid increased mass has been rarely described (Figure 1) [6, 13, 14].



Fig. 1: Congenital rapidly growing infiltrating lipoblastoma of the right forearm.

## III. PATHOLOGIC CHARACTERISTICS

**In gross pathologic features**, lipoblastoma may be gray-white rather than yellow of mature lipoma (Figure 2). The tumor has a gelatinous consistency. The circumscribed or focal lipoblastoma is well encapsulated, whereas lipoblastomatosis is infiltrative and lacks a capsule [1, 7, 9].



Fig. 2: Lateral radiograph of the forearm shows a large mass with prominent radiolucent fat. Note the thickening of the anterior cortex of the radius.

**In microscopic features**, lipoblastomas tend to appear as multiple lesions with a lobulated pattern [5, 8]. These lesions are composed of monovacuolated and multivacuolated lipoblast, spindled to stellate mesenchymal cells, a plexiform capillary network, myxoid stroma, and mature adipocytes organized into lobules separated by fibrous septa (Figure 3). The myxoid component is often most prominent in very young patients [7].



Fig. 3: Photograph of the excised gross specimen reveals a gray-white, soft mass with a distinct capsular margin and focal fibrous tissue septa.

The histological patterns in lipoblastoma and lipoblastomatosis are identical.

**Molecular features**, lipoblastoma and lipoblastomatosis commonly demonstrate translocations that involve 8q11-13 and that lead to rearrangement of the *plag1* gene [3, 7].

## IV. RADIOLOGIC FEATURES

The consistent radiologic feature of lipoblastoma, regardless of imaging modality, is the presence of fat within the lesion [9]. Plain radiographs, ultrasonography (US) and Magnetic resonance imaging (MRI) scans are the main modalities for evaluating patients with lipoblastoma [1, 2, 7]. CT imaging entails significant radiation, and it no longer used routinely.

**Plain radiographs** show a radiolucent soft tissue mass with radiolucency similar to subcutaneous fat (Figure 4). Cortical thickening of an underlying bone or bone erosion are rarely seen [4, 15].

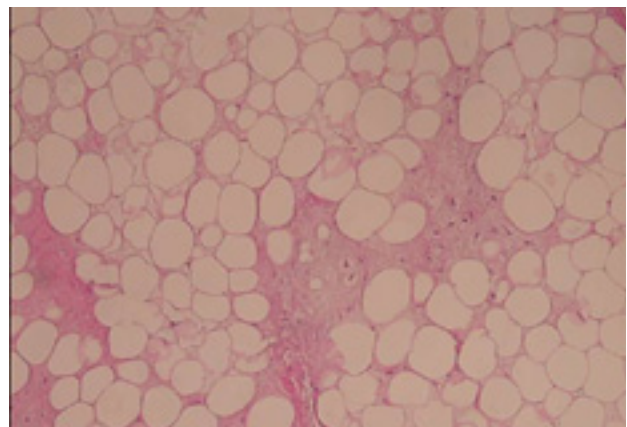
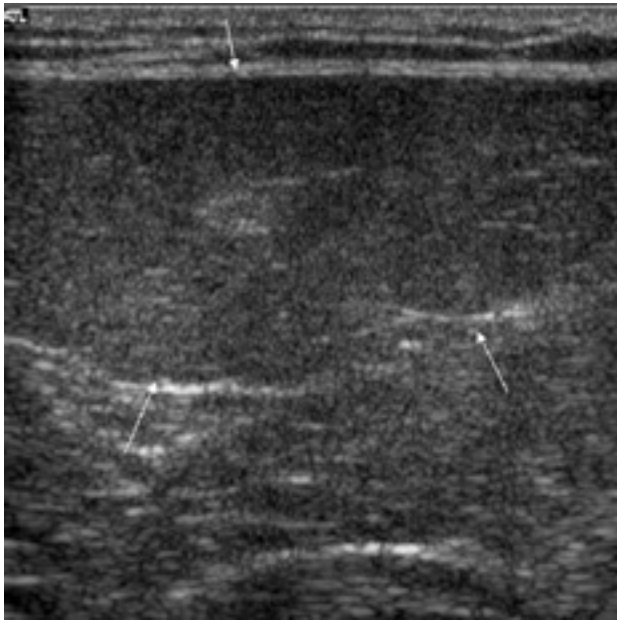


Fig. 4: Photomicrograph of lipoblastoma shows some extent of immature cells and lipocytes. Note the multivacuolated immature cells.

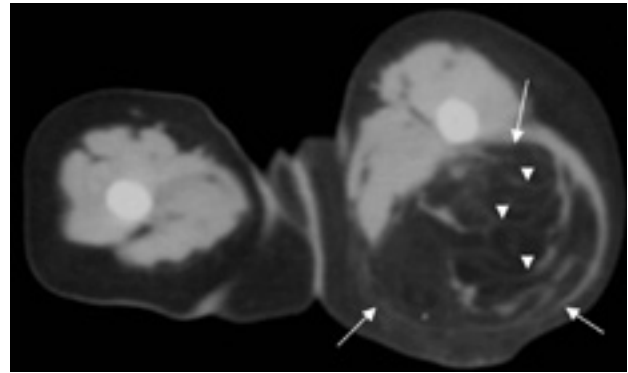
**At sonography**, a lipoblastoma typically appears as a homogeneously to finely textured, echogenic mass without detectable blood flow in color Doppler (Figure 5).



**Fig. 5:** Longitudinal sonographic image shows an elongated, well delineated and homogeneous hypoechoic mass (arrows).

Both CT and MR imaging can be used to confirm the presence of fat within the lesion.

**At CT imaging,** the lesion contains fat with low attenuation values (-60 to -120 UH), separate by septa of soft-tissue attenuation and do not enhance after administration of contrast material (Figure 6).



**Fig. 6:** Rapidly growing deep mass tumor 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 tumors had high signal intensity on MRI T1-weighted images and relative decreasing signal on T2-weighted images with signal loss on fat saturation pulse sequences (Figure 7a-c).

**At MR imaging,** lipoblastomas can be heterogeneous and have intermediate to high signal intensity on T1-weighted images according to the amount of immature lipoblasts [6, 16, 17]. Lipoblasts have lower signal than lipocytes on T1-weight sequences [18]. On fat-suppressed MR images, lipoblastomas usually demonstrate areas of high signal intensity, which can suggest the diagnosis. This finding may



**Fig. 7:** Intramuscular fatty tumor 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 tumor with complete absence of signal.

be of potential value in differentiating lipoblastomatous from lipomatous lesions and from exceptional infantile liposarcoma [6, 8, 19-21].

Lipoblastomatosis reveals similar intrinsic imaging features but also demonstrates infiltrative growth involving both subcutaneous tissue and muscle without a surrounding capsule (Figure 8 a-d).

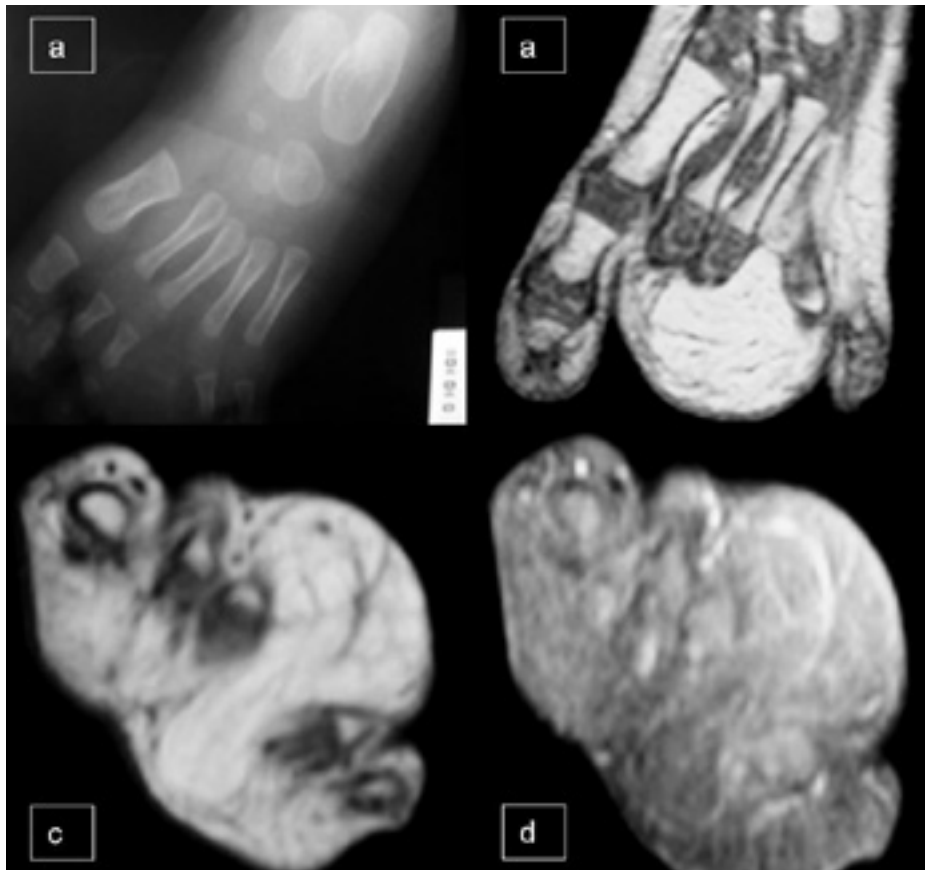
In very young patients (infants), the myxoid components may predominate with only small elements of fat. These myxoid areas are hypoechoic at sonography, low attenuation at CT, and at MR imaging are low signal intensity with T1-weighted sequences and high signal intensity with T2-weighted sequences, reflecting their high water content. These areas also enhance with contrast mate-

rial, owing to the rich capillary network. Lipoblastomas with this imaging appearance are indistinguishable from a myxoid liposarcoma. However, the age of the patient is vital in allowing accurate diagnosis [7].

## V. DIFFERENTIAL DIAGNOSES

Differential diagnoses include all fatty tumors in children, such as lipoma, lipomatosis, hemangioma, hibernoma and liposarcoma.

**Lipoma:** is a well-defined fatty tumor 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 [22].

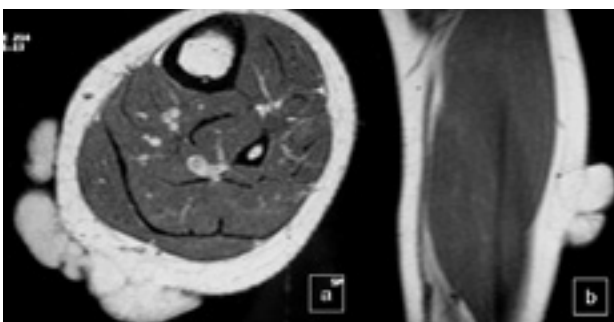


**Fig. 8:** Interdigital commissure well circumscribed lipoblastomatosis in a 16-month-old boy. a) Plain X ray of the left foot showing a 4th toe pushed back by a soft tissue mass. b) Axial T1-weighted shows a well defined hyperintense mass in contact with the 3rd toe and driving the 4th toe. c), d) Coronal T1-weighted and coronal T1 weighted with Fat Sat after gadolinium injection confirm fatty nature and show dorso-plantar mass with interdigital commissure infiltration.

This tumor 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.

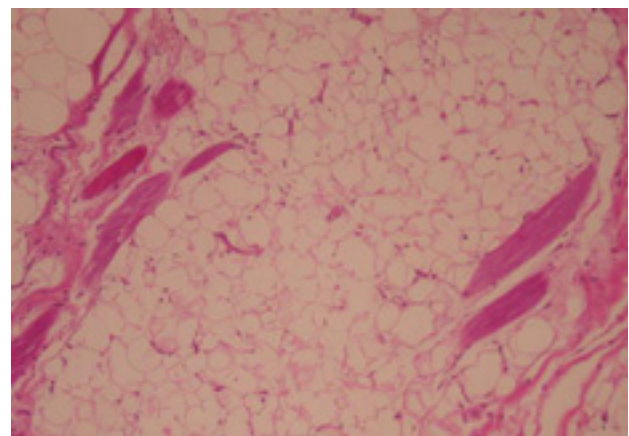
Ultrasound finds all its interest in these surface localizations, it makes it possible to establish the diagnosis by showing a lengthened circumscribed mass, whose large axis is often parallel with that of the affected limb, measuring typically less than 5cm, hyperechoic, non vascularised and depressing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

Ultrasound 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 tumor of soft tissues of the limbs" (Figure 9) [23].



**Fig. 9:** Superficial lipoma with an exophytic appearance of the left leg in an 11-year-old boy. a) Axial and b) Sagittal T1-weighted of the left leg showing a superficial lobulated mass hyperintense continuing the subcutaneous fat.

The histological examination confirms the diagnosis by showing a well limited tumor, lobulated consisting in mature adipocytes with conjonctivo-vascular septas (Figure 10).



**Fig. 10:** Histological section of lipoma. Note the mature fatty cells dissociating the striated muscle fibers.

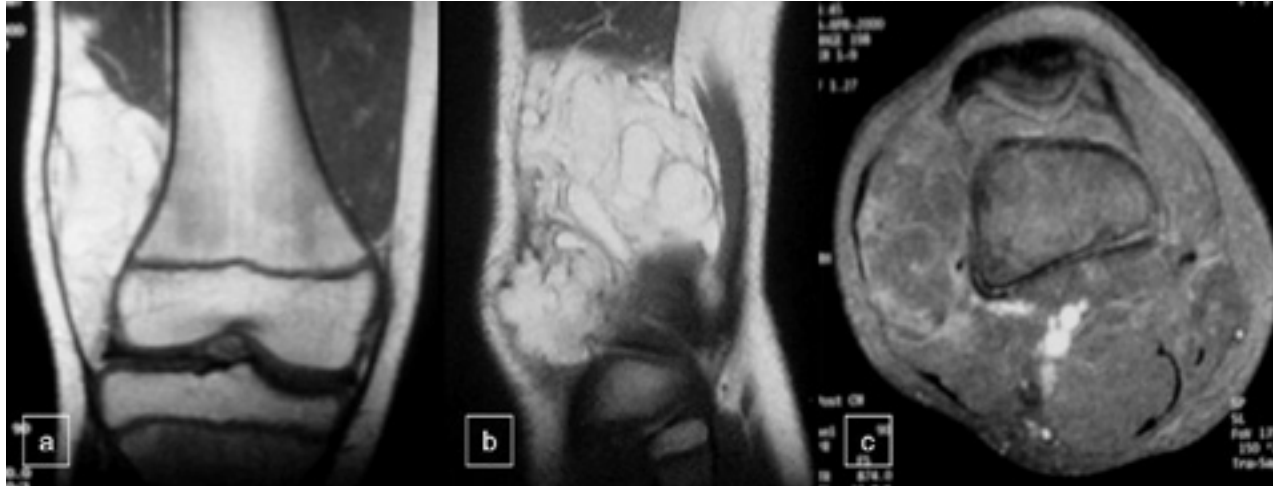
**Lipomatosis or infiltrating lipoma or infiltrating intramuscular lipomas:** are so named because they infiltrate skeletal muscle. These tumors are extremely rare in the child population [6, 24, 25]. They tend to be circumscribed, but non encapsulated. In children, these lesions most often appear by a muscular distortion or enlargement of an entire extremity or a palpable mass [24, 25]. They are more easily found on contraction of the involved muscle [5]. Impingement or invasion of nerves can result in sensory or motor deficit [6]. The most common sites are

large muscles of the extremities, especially those of the thigh, shoulder, and upper arm [5, 25, 26].

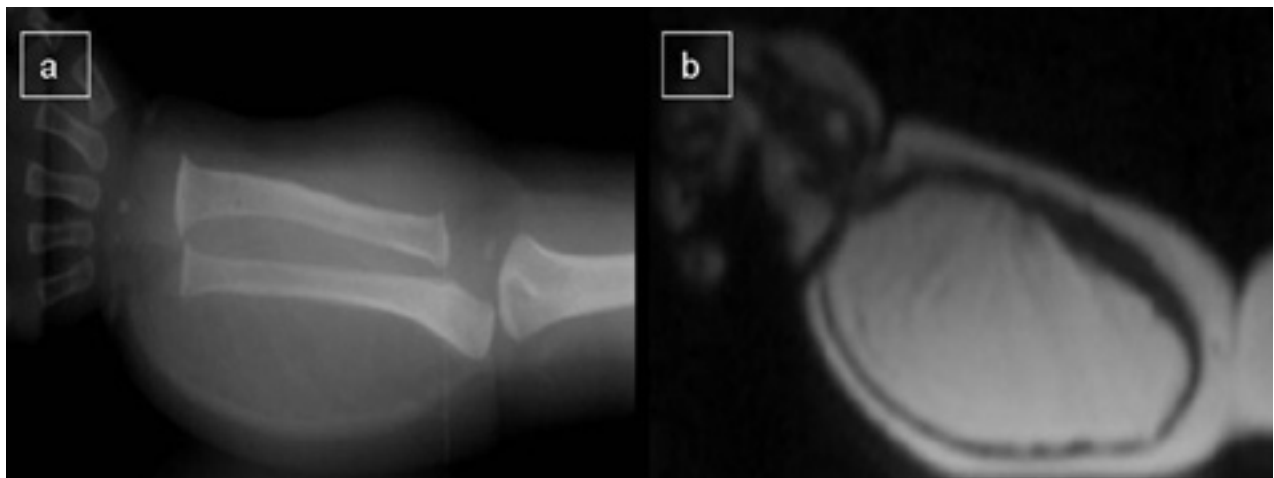
Plain radiographs show location in the muscular plane with sometimes a feathery appearance caused by strands of skeletal muscle running through the mass [25].

Intramuscular lipomas tend to be uninodular on MRI

scans and although infiltrative they tend not to destroy the normal tissue around them [20]. 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 (Figure 11, 12) [6].



**Fig. 11:** Deep painless soft tissue mass of the right knee had been present for several months in a 14-year-old boy. a), b) Coronal T1-weighted show a sub fascial hyper intense mass with thin fibrous septa adhere with the ilio-tibialis tractus. c) Axial T1-weighted with Fat Sat shows suppression of the signal intensity within the mass. This mass is relatively sharply delineated from the adjacent structures; however adhesion with lateral retinacular is noted making it resected while surgery. There was no bony involvement. The differential diagnosis should include intra muscular lipoma developed in the vastus lateralis. This mass was removed in its entirety and was histopathologically proven to be an intramuscular lipoma.



**Fig. 12:** 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 plan 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.

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 [5].

Complete surgical excision is recommended. Recurrence appears to be relating to an incomplete resection [5].

**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.

**Other benign soft tissue lipomatous lesions:** hemangioma, hibernoma, chondroid lipoma, angioliipoma and myoliipoma 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 which has signal intensity similar to that of fat on MRI with typical branching vascular structures [27].

On ultrasound, angioliipoma is echogenic 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.

**Liposarcoma** is a malignant tumor of mesenchymal origin which is extraordinarily rare in patients less than 10 years of age [1, 7]. Congenital liposarcoma are extremely

rare but have been reported [10]. This tumor is classified into the well differentiated, myxoid, round cell, pleomorphic and dedifferentiated types. Lipoblastoma shows close resemblance to low grade liposarcoma, especially the myxoid and the well-defined variant types [2, 8, 19, 20, 26, 28, 29]. The absence of any atypical lipoblasts, mitoses, hyperchromatic nuclei absence of atypia distinguishes lipoblastoma from liposarcoma. Further more, well-differentiated liposarcoma tend to be multinodular, more invasive and have a large amount of peripheral oedema. Septations tend to be thicker and enhance to a greater extent, because often there are muscle fibers within these septa [30, 31].

## VI. TREATMENT AND PROGNOSIS

Surgical excision is the treatment of choice and the prognosis is excellent. It is now well established that lipoblastoma is a benign tumor best treated by complete surgical excision and that a radical operation is not necessary and may be mutilating, especially in children [32]. 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 [5, 8, 14, 24, 32].

These tumors have an excellent prognosis despite their potential to invade locally and to grow rapidly to a considerable size [8, 32]. 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 [6, 33].

## VII. CONCLUSION

Lipoblastoma behaves benignly, occurs in both superficial and deep sites, and occasionally attains large size. Imaging findings are helpful in identifying the lipomatous nature of the mass. Complete surgical excision is the treatment of choice and long-term follow-up is required because there is a reported tendency for these tumors to recur.

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