Lipoblastoma: case report and literature review

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Abstract

Introduction: The term lipoblastoma was introduced by Jaffe in 1926. It is a benign mesenchymal neoplasm of uncommon embryonic adipose tissue, made up of adipocytes in different stages of maturation, 90% of cases occurring in patients younger than 3 years. **Case report:** The case of a 2-year-old patient with a tumor in the upper third of the right thigh is described, with poor adherence to medical surveillance, performing definitive treatment at 6 years of age with a histopathological diagnosis of lipoblastoma. **Discussion:** Lipoblastoma occurs most frequently in extremities and trunk, progressive growth; magnetic resonance is the most sensitive radiological study to make diagnosis, and treatment is surgical. **Conclusion:** Lipoblastoma is a benign neoplasm of soft, mesenchymal parts, which occurs predominantly in childhood; there is a risk of recurrence in both the localized and the diffuse form, consequently surveillance is desirable for 5 years after resection. Our patient is without recurrence and with good quality of life.

Key words: Lipoblastoma. Magnetic resonance. Surgical.

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Introduction

The term lipoblastoma was introduced by Jaffe in 1926, indicating that this tumor is made up by embryonic adipose cells; the largest published series is of 35 cases (Chung and Enzinger, 1973)^{1,2}. Lipoblastoma is a rare benign embryonic adipose tissue mesenchymal neoplasm constituted of adipocytes at different maturation stages that predominantly occurs during childhood^{3,4}.

According to Chung and Enzinger criteria, there are two clinical forms: a) lipoblastomas, which are circumscribed, subcutaneous neoplasms, with predominance in the limbs, and that account for 70% of cases; b) lipoblastomatosis, of infiltrative, diffuse form and deep localization⁴⁻⁶.

This lesion has an occurrence ratio of 3:1 according to gender, with predominance of the male gender, with 90% of cases occurring in patients younger than 3 years²⁻⁷. Seventy percent of localizations are at the upper and lower limbs, the trunk, the head and the nape; inguinal canal, intrathoracic and retroperitoneal localization has also been reported in the literature^{5,8,9}.

Case report

This is the case of a 2-year old female patient with no past medical, family or hereditary history relevant to the case, who attended for the first time owing to the presence of a mass at the right thigh upper third, with an incisional biopsy being performed and lipoma being diagnosed. Subsequently, follow-up was neglected by the family for 4 years, until reassessment at 6 years of age (Fig. 1).

Computed tomography (CT) showed an oval-shaped tumor lesion of solid component, surrounded by a fatty halo, with low contrast medium enhancement, localized at the right thigh posterior and anterior compartment; it pushed adjacent muscles (adductor longus, gracilis and sartorius) and neurovascular bundle at the femoral level mainly towards the anterior region (Fig. 2); in lower sections, the tumor lesion continued with the same characteristics, pushing the semimembranosus, semitendinosus and femoral biceps muscles towards the posteromedial zone.

The coronal reconstruction showed tumor total delimitation, completely surrounded by solid component (fat), and a focus of central necrosis. The sagittal reconstruction corroborated that the lesion was not dependent on bone tissue and that it was localized at the thigh anterior compartment (Fig. 3).

Figure 1. Six-year old patient with presence of tumor at right thigh upper third.

Figure 2. Tumor at posterior and anterior compartment, adjacent muscles pushed towards anterior region and neurovascular bundle at the femoral level.

Surgical procedure for wide resection was carried out on 04/02/2016, where using an inguinal approach with extension to medial compartment, a tumor with lipomatous appearance of approximately 20 cm in diameter was found, with the lesion pushing muscular planes, no data consistent with macroscopic infiltration and extending until being in touch with the maximus and minimus glutei posterior aspect with superior vascular pedicle. Resection was uneventfully carried out with preservation of vascular and nervous structures; bleeding was less than 100 mL and surgical time was 3 hours. The

Figure 3. Sagittal section showing bone tissue-independent lesion located at posterior compartment of thigh.

Figure 4. Nuclear positivity for S-100 protein (60x).

patient was subsequently surveilled, the Drenovac® was removed at 12 days and the stitches at 15 days.

Macroscopic examination showed a of 510 g specimen of 20 x 15 x 10 cm, with dark brown surface, vasculature firm in consistency, finely encapsulated. When sectioned, it revealed a 10 x 15 cm fibrosis area with pinkish tissue of myxoid consistency.

Microscopic examination of the histologic preparations under study identified a benign neoplasm of mesodermal

lineage, constituted of ovoid cells with no atypia and finely vacuolated cytoplasm with ill-defined borders, which react to anti-protein S-100 antibody (Fig. 4), which in turn is individually distributed on a myxoid stroma with irregular collagen fibers (Fig. 5). In other areas, the neoplasm displays adult-type adipose cell groups, with no atypia, which appear separated by thick septa of fibroconnective tissue (Fig. 6). Diagnostic impression: lipoblastoma.

A control CT showed postsurgical tumor bed with data consistent with fibrosis; most superior structures returned to their usual localization.

Currently, at one year of surgery and with no evidence of recurrence, there are no postsurgical sequels. Preserved muscular strength (5/5), preserved muscular tone and normal muscle stretch reflexes (++). With excellent quality of life.

Discussion

First contact with the patient was at 2 years of age, when she attended due to the presence of right lower limb tumor, the biopsy histopathological result of which indicated a lipoma; however, treatment was carried out at 6 years of age¹⁰. Considered an almost exclusively pediatric pathology and, in spite of the 3:1 gender-prevalence reported for males, our case occurred in a female. Lipoblastoma occurs more commonly in the limbs and trunk and has an average of 5 cm in diameter, but it can reach up to 20 cm^{5,6}. In most cases, the patient is asymptomatic until volume increase is detected or when any neurovascular or visceral structure is compromised^{2,4,5}.

Radiologic diagnosis includes MRI, the most sensitive test, which enables adequate definition of tumor limits and differentiation of compromised planes and structures, as well as of their components. Since it is of fatty origin, lipoblastoma is presented in basic sequences; at T1, it shows an intermediate signal secondary to immature lipoblasts, and at T2, it is hyperintense, heterogeneous and predominantly fatty. Although with less sensitivity, CT can also be useful for diagnosis, usually at -60 to -100 Hounsfield units, extremely hypodense structures are observed, which helps to determine if there is bone involvement, calcification or hemorrhage within the lesion¹⁻³.

Preoperative diagnosis is difficult owing to the tumor presentation size and because it is heterogeneous. Generally, it is mistaken with other soft-tissue tumors, both benign and malignant^{3,8}.

Figure 5. A: Myxoid stroma (alcian blue staining-positive, 60x). **B:** Lipoblasts and collagen fibers (hematoxylin-eosin staining, 60x).

Figure 6. Adipose cells (hematoxylin-eosin staining, 60x).

Treatment of these tumors is surgical, with complete resection (R0) being adequate; recurrence index ranges from 9 to 25%, with higher percentages occurring in patients with incomplete resection, generally within the 2 first years^{3,10}. Follow-up for 5 years is recommended^{6,9}. In none of the reviewed series has there metastatic disease been reported or has there any beneficial effect with chemotherapy, radiotherapy or corticoid treatment been demonstrated^{3,7}.

Lipoblastoma differential diagnosis should be made with other soft-tissue tumors such as liposarcoma, clear cell

sarcoma, synovial sarcoma, rhabdomyosarcoma, intramuscular lipomas, angiolipomas and hemangiomas^{2,11,12}.

Lipoblastomas show cells that are positive for vimentin and S-100 protein, and negative for other markers, including cytokeratin, CD34, desmin and NKIC3².

Genetic testing shows specific chromosomal anomalies with chromosomal rearrangements, *PLAG1* gene chromosome 8 (8q 11-13) deletion^{1,3,5}.

Conclusion

Lipoblastoma is a soft tissue, mesenchymatous benign neoplasm, with predominant occurrence in childhood; the only treatment known to this moment is surgical. There is risk of recurrence of both the localized and the diffuse forms, and surveillance for 5 years after resection is therefore advisable. The diagnosis can be suspected according to age group and radiologic characteristics.

In localized cases, surgical resection with anatomic structures preservation is able to preserve limb function and offers the patients a normal quality of life.

Conflict of interests

The authors declare there are no conflicts of interests relevant to this work.

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