Rapidly Growing Facial Tumor in a 5-Year-Old Girl

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Abstract
The aim of this study is to describe the clinical, radiological and histological characteristics that define lipoblastomas with special emphasis on differential diagnosis. The patient is a 5-year-old girl who consulted for a rapidly growing lower cheek tumor. This study analyzes, evaluates, and discusses the issues that need to be addressed throughout the process that affect treatment planning and provides an updated review of these rare head-and-neck tumors.

Keywords: Head-and-neck tumors, lipoblastoma, lipoblastomatosis, pediatric age

INTRODUCTION
Lipoblastomas are very rare benign tumors that occur in early childhood.[1-3] They constitute from 4% to 6% of soft-tissue tumors in children, with a slight male predominance.[1-3] They arise from embryonic adipose tissue and are characterized by rapid growth, with symptoms depending on their location, and can cause deformity or compression of adjacent structures.[1,4]

They are commonly found in the trunk and extremities and are uncommon in the head and neck.[1-4] There are two clinical forms with similar histology: lipoblastoma, a localized, superficial, and encapsulated form accounting for 70% of the cases, and lipoblastomatosis, a diffuse, deep form, usually infiltrating adjacent muscle tissues.[1-3]

The differential diagnosis of these tumors can be complex, ranging from inflammatory lesions, infectious causes, traumatic injuries, and vascular or lymphatic malformations to other benign tumors such as lipomas or hibernomas and malignant tumors such as myxoid liposarcoma.[1-5] Suspected diagnosis should be based on clinical and radiological data, especially magnetic resonance imaging (MRI), which is considered the test of choice as it provides high-quality images without radiation;[1,6,7] however, confirmatory diagnosis is always histological.[1,8] The most accepted management for these lesions is complete but nonmutilating excision, which has a very good long-term prognosis.[1,6]

We evaluated all case characteristics, focusing on clinical and radiological data essential for the maxillofacial surgeon who must remember two aspects when considering an appropriate differential diagnosis: first, these tumors are very rare in the head and neck and almost only affect children; second, they can be very difficult to distinguish from malignant lesions such as liposarcoma. An updated review of the literature is also provided.

CASE REPORT
A 5-year-old girl without a relevant history was referred from the pediatric department to study a rapidly growing facial tumor; the initial suspected diagnosis was cellulitis of dental origin because of its location.

Physical examination presented a soft mass with 2 months of evolution in the right lower cheek, overlying the mandible.

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Movable, nonadherent, nonpainful, well-circumscribed mass on the right mandibular border without local signs of infection, and with rapid growth causing noticeable facial deformity [Figure 1]. The size of the mass was approximately 3 cm × 2 cm, and no murmurs or pulsation was detected in the tumor. Clinically, the patient was asymptomatic at all times, without fever or pain, and there was no difficulty in chewing or swallowing. Examination of the oral cavity was normal.

An imaging study was performed using orthopantomography, which was normal [Figure 2], and MRI (Systems Achieva Release 3.2 Level 3, 1.5 T, Philips Medical Systems, the Netherlands), which revealed a well-delimited homogeneous lesion (3.9 cm × 1.6 cm maximum diameter) adjacent to the right mandibular cortex displacing the anterior edge of the masseter without infiltration; the lesion appeared hyperintense on T1- and T2-weighted sequences, which became hypointense in fat-suppressed T1 and T2 sequences, showing the same signal intensity as fat in all pulse sequences [Figures 3a, b and 4a, b]. Considering these radiological characteristics, the initial suspected diagnosis was an adipose tissue tumor; in the differential diagnosis, we included lipoma, lipoblastoma and, less likely, liposarcoma.

After a thorough study of the case using clinical and radiological data, lipoblastoma was considered the most likely diagnosis, opting for complete excision of the lesion. For this purpose, an intraoral approach was used to avoid external scars and damage of the marginal branch of the facial nerve, carefully preserving the exit of the mental nerve. The lower cheek musculature was detached, and the tumor was resected. After complete excision, an encapsulated, lobulated lesion of bright, yellowish appearance was observed macroscopically with regular, lobulated, and smooth borders [Figure 5a and b].

Histology confirmed the diagnosis of lipoblastoma, revealing a lesion with a predominance of mature and immature adipose tissue, areas of slight fibroblastic proliferation without atypia, myxoid stroma, and the presence of vessels [Figure 6a and b]. The postoperative course was uneventful, and no paresis of the facial nerve was observed. At 1-year follow-up, the patient showed a good facial symmetry and no recurrence [Figures 7 and 8a, b].

**DISCUSSION**

Lipoblastoma is a very rare tumor of embryonic fat cells, characteristic of children under 3 years of age, accounting for
90% of the cases described.\cite{4,9,10} These tumors are very rare in the head and neck. A review conducted by Dutton et al.,\cite{11} which includes a series described in literature until 2011, reports on 411 cases, of which 68% were lipoblastomas and 32% were lipoblastomatosis; of these, only 17% were in the head and neck; 54 cases in the neck and 19 cases in the face and head. This review is updated to the present time, but only for lesions in the head and neck, reporting 88 cases of lipoblastoma and 31 cases of lipoblastomatosis in the head and neck; approximately 65% of the cases involved the neck and only one the lower cheek overlying the mandible [Table 1], two cases if we add the case reported here.

The pathogenesis is unknown, though they are believed to arise from altered embryogenesis of human white fat and genetic predisposition, as chromosome 8 abnormalities may contribute to the development of lipoblastoma.\cite{5,9,11} The diagnosis of these lesions is only simple if there is an appropriate clinical context and MRI confirms a fatty lesion.\cite{5,6,9,29} Although there are no pathognomonic signs of lipoblastoma, some fundamental aspects must be considered, such as clinical characteristics of the lesion (soft, movable, nonpainful, rapid growth, almost exclusive to children, and rarely produces symptoms until growth, which can cause deformity or compression symptoms resulting in swallowing difficulty or even Horner’s syndrome)\cite{1,5,7,9,25} and diagnostic imaging, specifically MRI, which is considered the test of choice to determine a diagnosis because it allows confirmation of an adipose tissue tumor and its relationship with adjacent structures and is useful for follow-up and early diagnosis of recurrence.\cite{5,6,9,29} Radiologically, these lesions show fat density in all sequences, revealing high-intensity masses in T1- and T2-weighted sequences and hypointense masses in fat-suppressed sequences, usually without peripheral infiltration, except for lipoblastomatosis infiltrating muscle.\cite{1,4,7,9}

After a thorough study of clinical and radiological data, the differential diagnosis should be limited to lesions showing similar characteristics such as lipoma, hibernoma, and malignant lesions such as liposarcoma.\cite{1,2,4,9}

Confirmatory diagnosis is always histological; lipoblastoma is characterized by proliferating embryonic white adipose tissue with multivacuolated lipoblasts, myxoid stroma, plexiform vessels, and cellular pleomorphism. However, these characteristics can be ambiguous and overlap with other lesions such as simple lipoma, hibernoma, and, importantly, malignant lesions such as myxoid liposarcoma.\cite{1,4,8,15,29}

Lipoma is common in adults, very rare in children, and usually grows slowly; it is histologically characterized by mature fat cells and the absence of lipoblast, rarely showing lobulation.\cite{2,8} Hibernoma is also characteristic of adults, is very rare in children under 18 years of age, and is histologically defined as a tumor of embryonic brown fat, showing characteristic
eosinophilia; also, it usually has a lobular pattern and granular cytoplasm not present in the white fat of lipoblastoma.\textsuperscript{[2,11,15]} Regardless, both lesions are benign, and the treatment is complete excision.\textsuperscript{[2,8,11,15]}

Issues arise when there are doubts regarding malignant lesions such as myxoid liposarcoma, which, despite being a very rare neoplasm (5% of soft-tissue sarcomas), is the most commonly described tumor in children; it can have similar rapid growth characteristics; however, it is more common after 15 years of age, especially in the third decade of life.\textsuperscript{[1,4,6]} Histologically, in these cases, there may be lipoblasts in different stages of maturation, a plexiform capillary network, and a myxoid matrix, all suggesting lipoblastoma. However, liposarcomas also show nuclear atypia and hyperchromasia, absent in lipoblastoma, which are important distinguishing factors. Given the importance of the diagnosis of liposarcoma, if doubts persist, it is necessary toCapabilities of the system include, but are not limited to: reading and understanding natural language, generating responses in a coherent and fluent manner, and providing information in a clear and concise format.
perform an immunohistochemical and specific cytogenetic study to confirm the differences.[1,4,6,10,22]

Cytogenetic analysis reveals some features of lipoblastoma such as chromosomal abnormalities in adipose tissues of chromosomal region 8q11–13 and gene rearrangement involving the PLAG protein. In contrast, in myxoid liposarcoma, translocation t(12;16) (q13;p11) and FUS-DDIT3 gene fusion are characteristic in 95% of cases.[11,12,22,23] The presence of p16, useful for the diagnosis of liposarcomas, is highlighted in an immunohistochemical study.[25] With a well-founded suspected diagnosis, treatment consisting of complete but nonmutilating excision with free margins can be performed.[3,4,11] Prognosis is excellent, with a recurrence rate of 0%–25% related to incomplete resection of large lesions or muscle infiltration.[1,4,11] Although lipoblastoma is a benign lesion and no cases of malignant transformation have been recorded, a minimum 5-year follow-up is recommended.[1,3,4,7]

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient’s guardian has given consent for the child’s images and other clinical information to be reported in the journal. The patient’s guardian understands that the child’s name and initial will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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