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Observation

Rare Entity of Cervical Tumors: Lipoblastoma

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Abstract

Lipoblastoma is a benign tumor of white fat, of embryonic origin. It occurs exclusively in the small child, mainly at the trunk and extremities. Cervical localization is rare. It is of good prognosis after complete excision (WHO2013). We report the observation of cervical lipoblastoma collected at the department of pathological anatomy at Mohammed VI Hospital, Marrakech. It was a two-year-old infant. He had an ulcerated left cervical mass. On clinical examination, this mass was mobile with respect to the deep plane. Surgical excision was performed. On macroscopic examination, there are two well-defined, encapsulated, whitish, soft-like formations and seat of myxoid recessions. Microscopic examination shows a benign mesenchymal proliferation, arranged in lobules of variable size. The latter are delimited by fine irregular conjunctiva-vascular trabeculae. The lobules consist of lipoblasts at different stages of maturation, provided with oval nuclei, discreetly hyperchromic, without abnormal mitosis. The cytoplasm is microvacuolar and eosinophilic. Presence on the periphery of loose myxoid zones. The immunohistochinic study carried out objectified labeling the tumor cells with anti-CD34 and anti-PS100 antibodies with a Ki67 proliferation index of 1%. Lipoblastoma is a rare tumor of embryonic fat cells. This tumor primarily affects children under 5 years. There is a male predominance with a sex ratio of 2. Cervical localization is rare. The positive diagnosis is pathological. The differential diagnosis arises with liposarcoma in its myxoid variety. He has a good prognosis. It has a potential for local extension but does not metastasize. Excision surgery is the only treatment to prevent local recurrence.

Introduction

Lipoblastoma is a benign tumor of white fat, of embryonic origin. It occurs exclusively in the small child, mainly at the trunk and extremities. Cervical localization is rare. It is of good prognosis after complete excision (WHO2013).

Material and methods

We report the observation of cervical lipoblastoma collected at the department of pathological anatomy at Mohammed VI Hospital, Marrakech

Observation

It was a two-year-old infant with a sibling of 2, from a non-consanguineous marriage, well vaccinated, followed by ophthalmology for convergent strabismus who had a left upper cervical mass (sub-angulomaxillary) left. gradually increasing in volume and evolving in a context of general state conservation for 1 year. On clinical examination, this mass was of a well-defined soft consistency, mobile with respect to the 2 planes without inflammatory signs, endopharyngeal expression and associated ADP. The patient had a cervical CT showing a lobulated mass suggestive of cystic lymphangioma. Careful surgical excision was performed after identification and preservation of the rising branch of the facial nerve, vaque and spinal nerves and the internal jugular vein. On macroscopic examination, there are two well-defined, encapsulated, whitish, soft-like formations and seat of myxoid recessions (Figures 1).

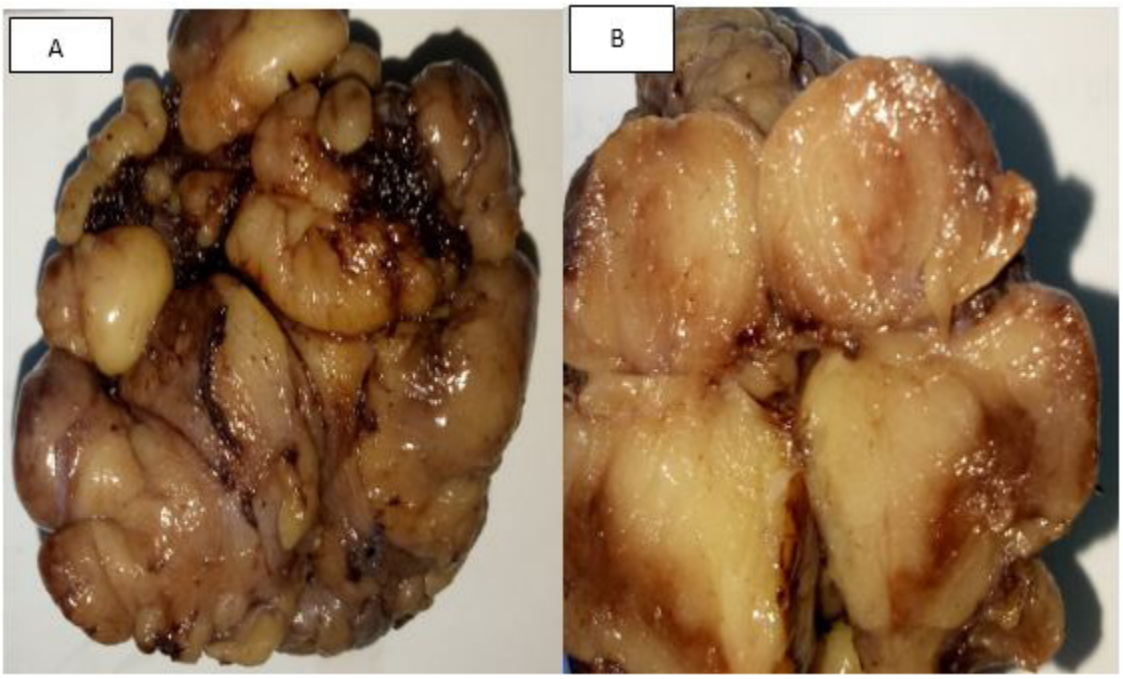

Figure 1: are two well-defined, encapsulated, whitish, soft-like formations and seat of myxoid.

Microscopic examination shows a benign mesenchymal proliferation, arranged in lobules of variable size. The latter are delimited by fine irregular conjunctiva-vascular trabeculae. The lobules consist of lipoblasts at different stages of maturation, provided with oval nuclei, discreetly hyperchromic, without abnormal mitosis. The cytoplasm is microvacuolar and eosinophilic. Presence on the periphery of loose myxoid zones (Figures 2).

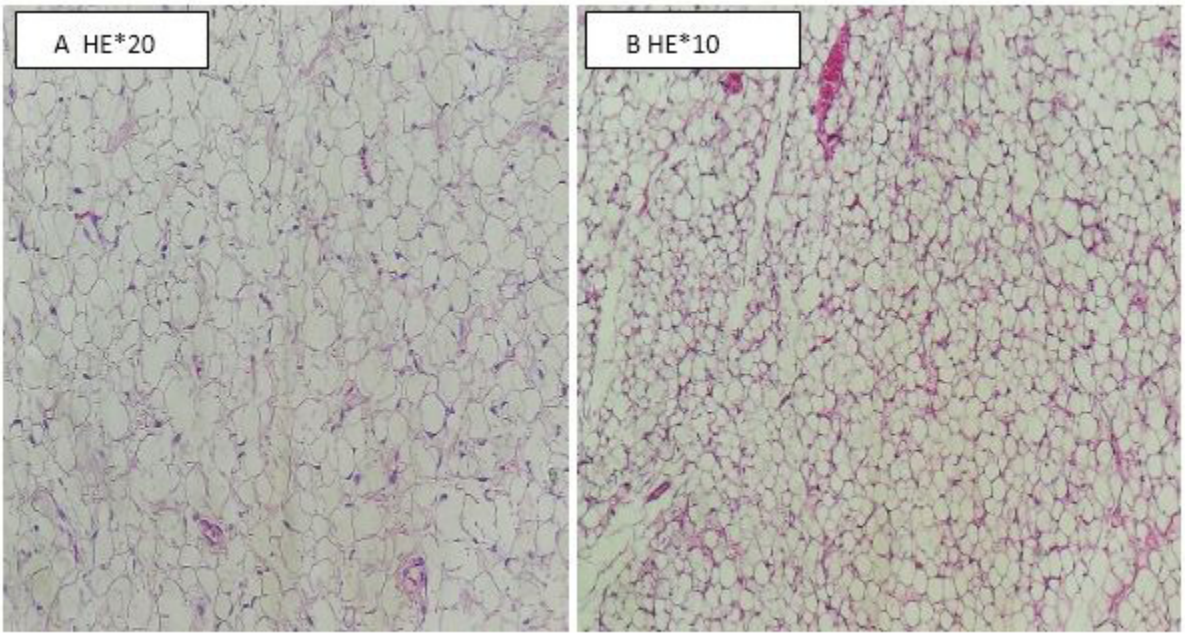

Figure 2: benign mesenchymal proliferation, arranged in lobules of variable size.

The immunohistochinic study carried out objectified labeling the tumor cells with anti-CD34 and anti-PS100 antibodies with a Ki67 proliferation index of 1%.

Discussion

Lipoblastoma is a very rare benign tumor of embryonic fatty tissue [1]. It is a tumor that affects almost exclusively the infant and the child. The age of onset is less than 3 years in 90% of cases and less than one year in 40% of cases [2]. Although rare, cases occurring in adults have been reported. The term lipoblastoma is introduced by Jaffe in 1926 [3]. In 1973, Chung and Enzingar [4] identified two forms: Lipoblastoma, a lobulated tumor corresponding to the circumscribed form of the tumor and lipoblastomatosis, a deeper, non-encapsulated and more infiltrating mass corresponding to the neighborhood structures corresponding to the form. broadcasts. The majority of these tumors affect the subcutaneous tissues of the extremities and the trunk. They are extremely rare in the head and neck: Only 48 cases have been reported in the English literature [5]. Lipoblastoma is usually asymptomatic, but it can become so by its size and location [6]. Rasmussen, et al. Mentioned a case of cervical lipoblastoma causing intermittent airway obstruction. San et al reported the first case of hemiparesis resulting from supraclavicular lipoblastoma by compression of the spinal cord [7].

Lipoblastoma presents itself clinically as a painless mass with slow growth reaching mainly the extremities (foot, hand). Very atypical localizations are however described (lip, pleura, mesentery). A mesenteric localization measuring 20 cm and weighing 450 g has been reported [8]. Lipoblastoma is never accompanied by secondary localization. It can, however, reoffend, especially if the excision is incomplete. For some authors however [9], there are cases of spontaneous resolution of this tumor, excellent prognosis and a conservative attitude may be preferable to a sometimes-mutilating surgery. Ultrasonography is often the first imaging performed in front of a tumor of the soft parts of the child because of its non-radiating, non-invasive character. It allows a first approach to tumor characterization: solid or liquid character, degree of vascularization. However, the etiological orientation and the extension assessment of the lesion remain limited. MRI is then often preferred [10] to the scanner to make a more precise assessment of the anatomical relationships of the tumor [10]. It allows the analysis in the 3 planes of the space of the relations of the tumor with the vasculo-nerve structures without irradiation. Lipoblastoma is typically 2-5 cm in diameter, although it can exceed 10 cm. The soft, tabulated, yellow, white, or tan mass may display myxoid nodules, cystic spaces, or fal nodules separated by fine white fibrous trabeculae.

Lipoblastoma characteristically demonstrates a tabular architecture with sheets of adipocytes separated by fibrovascular septa. Myxoid areas display a plexiform vascular pattern with primitive mesenchymal cells. The fat cells show a spectrum of maturation, ranging from primitive stellate or spindled mesenchymal cells, to multivacuolated or small signet ring lipoblasts, to mature adipocytes. The proportion of these cell types varies from case to case and from lobule to lobule. The fat lobule itself can exhibit a zonal pattern of maturation, with more immature myxoid cells at the periphery and mature adipocytes in the centre, although this pattern is not always maintained. Mast cells are common, Other histological findings include fibroblastic proliferation with collagen deposition, chondroid metaplasia, extram edullary haematopoiesis. chronic inflammation, and sparse multinucleated floret cells. Mitoses are very rare, and abnormal mitoses are absent. A lipomatous or fibrolipomatous pattern is seen as a manifestation of differentiation or maturation, with variable residual lipoblasts in myxoid zones (OMS 2013). On the morphological level, the architectural features observed in our observation correspond to those usually described [11]. They associate the well-defined [10] and homogeneous nature of the lesion, the presence of lobulations and fibrous septas. When tumor growth is rapid, there is the problem of differential diagnosis with liposarcoma especially in its myxoid variety with very poor prognosis. It is characterized histologically by the presence of abnormal mitoses [12,13] the lipoma that theoretically does not exist in children; hibernoma: a benign fatty tumor mainly affecting the young adult, electively affecting the thigh whose pathogenesis and potential for malignancy are still poorly understood [14]. The imaging aspect of these tumors is nonspecific. Histology offers a more precise approach to diagnosis. The detection of lobulated lipoblasts and the rarity of mitoses suggest the diagnosis of lipoblastoma [15]. The absence of abnormal mitosis rules out the diagnosis of liposarcoma. The hibernoma [14] has pathognomonic cytological characteristics (brown, uniform to small cytoplasmic mature fat cells).

The adipocytes of lipoblastoma demonstrate reactivity for S100 protein and CD34, and the primitive mesenchyma cells are often reactive for desmin (OMS). The cytogenetic study provides additional information for morphological analysis particularly useful for the diagnosis of lipoblastoma [16]. It often exhibits caryotipic rearrangements on chromosome 8 (fracture in the long arm) [12,13].

Conclusion

Lipoblastoma is a benign tumor of white fat, of embryonic origin. The clinical symptomatology has no specificity. The diagnostic certainty remains anatomo-pathological, the complete surgical excision makes it possible to avoid the recurrences which are frequent. Rigorous monitoring of at least three years must be undertaken.

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