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Original Research Article

Giant Cell Fibroblastoma in a 4 Years Old Child: Another Rare Case Report

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Abstract

Giant cell fibroblastoma (GCF) is a rare soft tissue most often discovered during the first two decades of life. We report the case of a 4-month-old infant examined for a mass in his right arm. The patient underwent total excision of the mass. Histological, immuno-histochemical showed giant cell fibroelastoma. This tumor poses diagnostic challenges to the pathologist because it may be confused with malignant mesenchymal tumors with different prognosis.

Keywords: Giant cell fibroblastoma, infant, rare.

INTRODUCTION

Giant cell fibroblastoma (GCFB) were first described by SHMOOKLER and ENZINGER in 1982[1]. It's a rare intermediate malignancy tumor, of childhood and young adults. This neoplasm can mimic clinically and even histologically the dermatofibrosarcoma protubrerans (DFSP), and is considered initially as the pediatric variant of this sarcoma [1,2-8]. the two lesions could be distinguished histologically and has been reported to co-occur together in the same tumor [2-12].

CASE PRESENTATION

Herein we report the case of a 4 years old boy presented to plastic surgery with a history of recurrent of the knee over the following three years. the mass progressively enlarged to 3cm.at the clinical examination, the patient was asymptomatic and free from any other lesions. Local examination found an infiltrative borders-lesion involving subcutaneous tissue, extending to superficial dermis. Surgical excision with free margins made by surgeons and the

Sample sent to our départements

Grossly it was a grayish white gelatinous consistence. Microscopically, it was a tumoral proliferation of spindle cells extending to the dermis and hypodermis. The spindle cells were arranged diffusely, with elongated nuclei and vesicular to hyperchromatic chromatin, eosinophile or scanty cytoplasm. The giant cells were scattered throughout the tumor containing variable number of round nuclei and abundant cytoplasm with irregular contours [figure 1]. pseudo vascular spaces were identified and they're considered characteristic of this tumor. Mitotic figures were rare with no vascular invasion or necrosis.

Throughout the tumor no mitosis or necrosis seen

Immunohistochemically, spindle cells were CD34 positive and some giant cells too. The spindle cells showed some positivity for PS100 too. [figure 2]

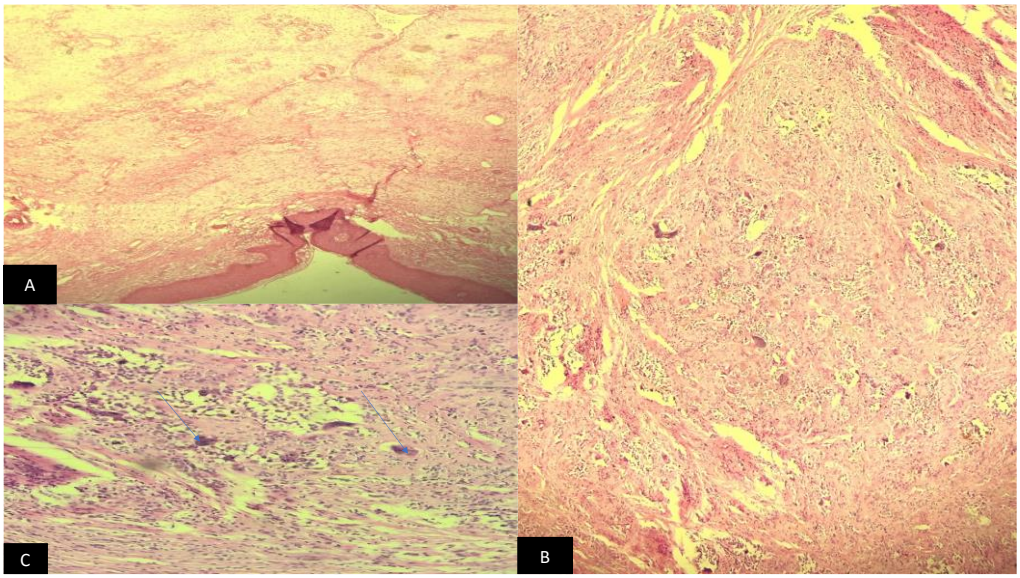

Figure 1: Hematoxylin-eosin stain [HE]. A: low magnification power[x10] showing tumoral proliferation of spindle cells extending to the dermis and hypodermis with some elongated dilated vascular spaces. B: med power magnification[x20] showing proliferation of spindle arranged diffusely, with elongated nuclei and vesicular to hyperchromatic chromatin, eosinophile or scanty cytoplasm, associated with numerous giant cells. C: high power magnification[x40]. cytological details; showing giant cells were scattered throughout the tumor containing variable number of round nuclei and abundant cytoplasm with irregular contours scattered

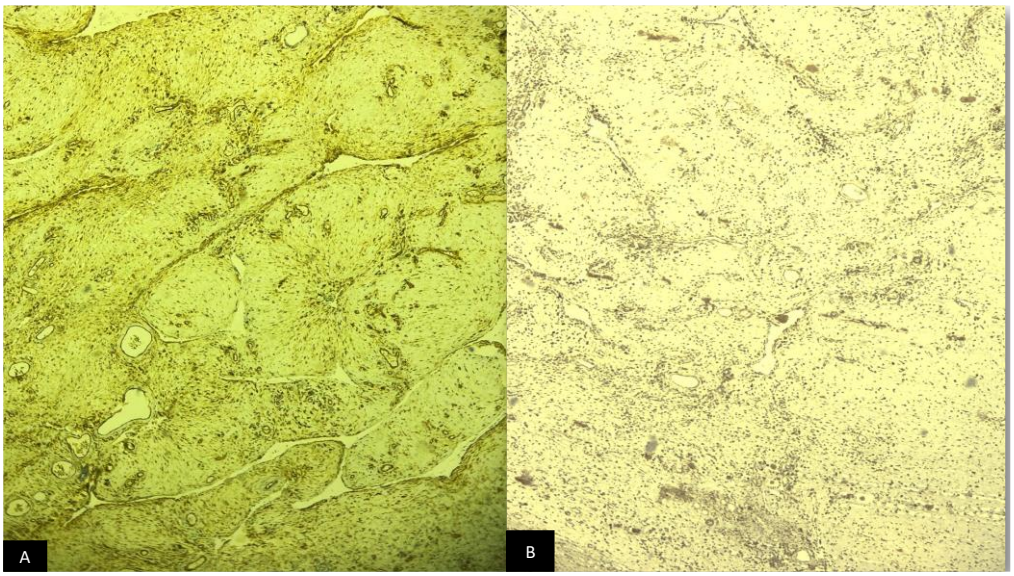

Figure 2: Immunohistochemistry analysis showing; expression of CD34 by spindle cells (A)and some giant cells. (B)the PS100 protein was weakly and patchy positive in our case

DISCUSSION

The GCFB is a rare tumor first described by smoker and Enfinger in 1982[1]. It generally occurs, almost exclusively, in the first two decades of life [2,3-6]. Less than 100 cases of CGFB have been reported [2,7]. thus, make believe that examples of this tumor were diagnosed as a low-grade sarcoma in the past. GCFB have predilection for thigh, inguinal region, and chest wall [8].it occurs in the soft tissue with possible extension to subcutaneous region. This tumor may be locally aggressive, but no metastases have been reported till now [9,10]. the gross presentation is the same; unencapsulated grayish gelatinous mass. histologically, CFB is characterized by infiltration of the mid and the lower dermis with bland spindle cells and multinucleated giant cells dispersed in a fibro myxoid stroma. mitotic activity is very low. A distinctive histological feature usually easy to observe, is the irregular pseudo vascular spaces lined by discontinuous row of multinucleated cells. the solid areas contain elongated to stellate cells within a stroma of variable cellularity.

Immunohistochemistry does not provide specific tools for diagnosis: the only antigen found

consistently GCFB is vimentin [11-16], CD34 is very useful as it's always expressed by the spindle cells as the giant cells sometimes, our case line this fact [12].

I histology, there is differential diagnosis of CFGB that should be taken in mind; angiosarcoma, myxoid liposarcoma, infantile my fibromatosis, lymphangioma. `but these entities are rare in the pediatric group. The tightest differential diagnosis is the DFSP as they share several histological features; both characterized by proliferation of spindle and stellate shaped cells within the dermis and with infiltration of subcutaneous tissue [7,8,17,18]. Immunohistochemically, they both express CD34 antigen [10]. there is published cases literature of association of the two neoplasms [7,8]. Furthermore, GCFB and DFSP present both the same molecular signature; the reciprocal translocation [17;22] [3,7,8].

In conclusion, GCFB is a rare soft tissue in pediatric group.it s considered as the pediatric variant of DFSP sharing similar histological and molecular features, and must kept in mind

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