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Giant Cell Fibroblastoma: A Case Report

Trabelsi F[1], Maamatou W[1*], Abid S[1], Daib A[1], Jabloun A[1], Ben Abdallah R[1], Hellel Y[1], Gharbi Y[1], Kaabar N[1]

Paediatric surgery department, Habib Thameur Hospital, Tunis, Tunisia[1]

Corresponding Author: 1*

ABSTRACT— Giant cell fibroblastoma (GCF) is an uncommon, mesenchymal neoplasm of the soft tissues. It is an unusual childhood tumour of intermediate malignancy. We are reporting a case of giant cell fibroblastoma in a 3 and a half-year-old boy who presented with painless oval swelling at the left shoulder that appeared from birth. MRI of the shoulder revealed a subcutaneous mass. The patient underwent a total excision of the mass. Histological, immunohistochemical and genetic examinations have shown giant cell fibroblastoma. This tumour poses diagnostic challenges to the pathologist because it may be confused with malignant mesenchymal tumours with different prognosis.

KEYWORDS: Giant cell fibroblastoma, childhood tumour, intermediate malignancy, surgery

1. INTRODUCTION

Giant cell fibroblastoma (GCF) is an uncommon, mesenchymal neoplasm of the soft tissues. It is an unusual childhood tumor of intermediate malignancy. It was originally described by Schmookler and Enzinger in 1982 in a series of twenty cases [1]. Giant cell fibroblastoma characteristically occurs in males during the first decade of life, with more than half of cases seen before the age of 5 years [2]. Giant cell fibroblastoma merits attentive interpretation as they have been misdiagnosed as sarcoma in the past leading. We are hereby reporting a case of giant cell fibroblastoma in a 3 and a half-year-old boy who presented with painless oval swelling at the left shoulder that appeared from birth.

2. Case Report

A 3 and a half-year-old boy presented with painless oval swelling at the left shoulder that appeared from birth with very significant increase in size for the last 6 months. Initially, the swelling was small and gradually increased to the present size of 8 × 8 cm. There was no history of trauma, fever, restriction of movements or any other swelling. On local examination, a freely mobile swelling of 8 × 8 cm was present at the left shoulder. It was soft, painless, and purplish and without local inflammatory signs (Fig. 1). An initial diagnosis of haemangioma of the left shoulder was made on radiological explorations (ultrasound, MRI) (Fig. 2). A surgical treatment of the tumour is then indicated. The intraoperative aspect was suspected of malignancy in front of the notion of the non-encapsulated mass with invasion of the surrounding tissues (skin and muscle). An extemporaneous examination concluded with a malignant tumour reminding a rhabdomyosarcoma. A wide excision of the tumour was done in addition to the cutaneous part which appears invaded. Gross specimen comprised a single, well-circumscribed, skin-covered, creamish white tissue piece measuring 9 ×7× 2 cm and appeared gelatinous on cut section (Fig.3). The final histological examination concluded with a giant cell fibroblastoma with complete excision. The patient was regularly followed up. A control ultrasound, three months later, did not show any nodules or tumour residue.

3. Figures

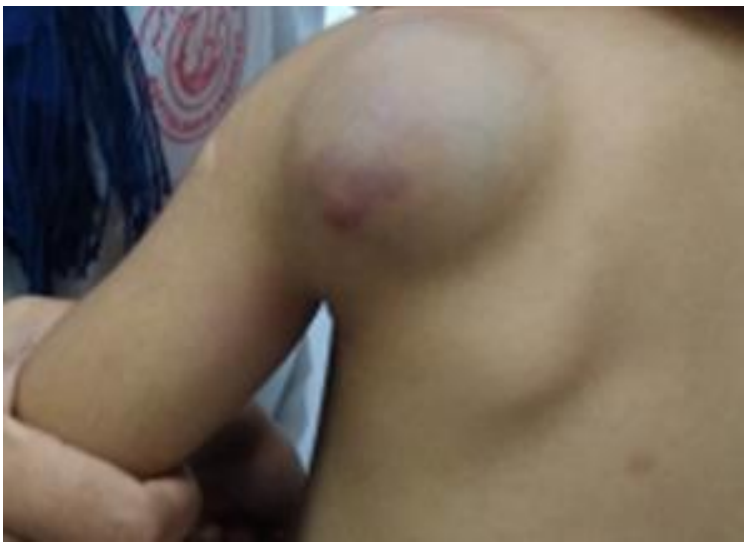

Figure1: Swelling of the left shoulder, purplish, without local inflammatory signs measuring 8 * 8cm

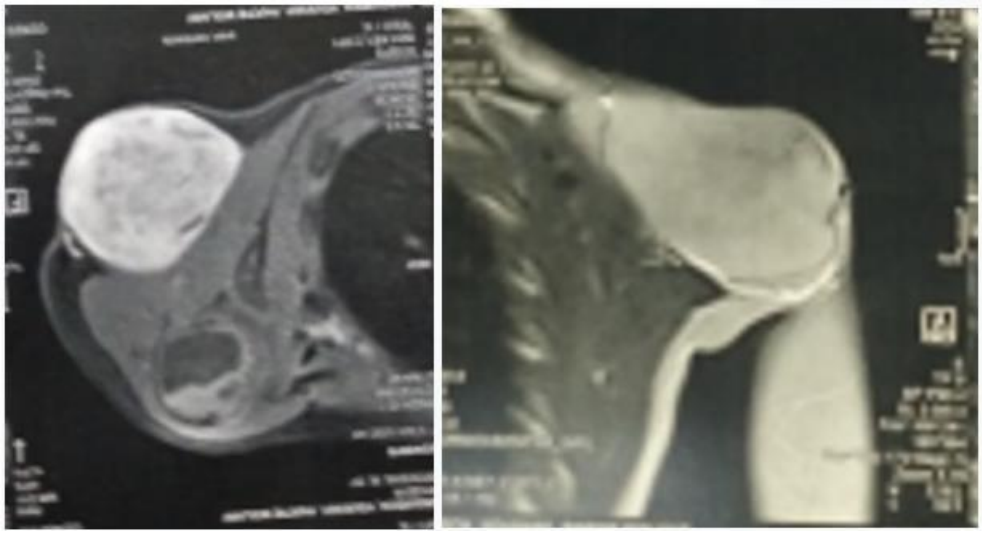

Figure 2: Haemangioma of the left shoulder on MRI

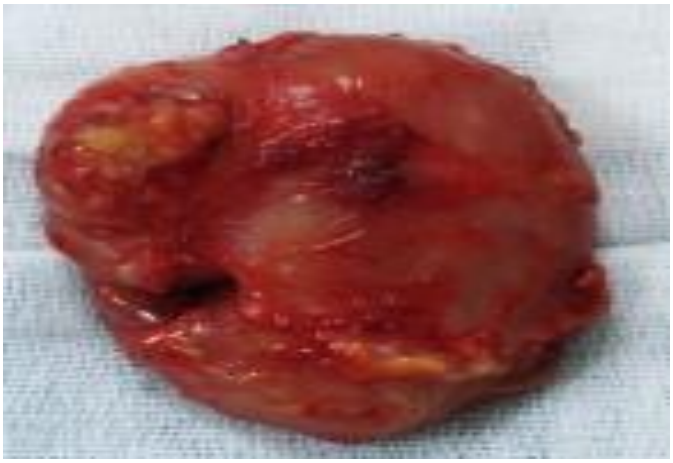

Figure 3: Gross cut section of skin covered well circumscribed creamish white gelatinous tissue piece.

4. Discussion

Giant cell fibroblastoma (GCF) is an intermediate grade, soft tissue tumour of childhood with high incidence of local recurrence and is seen in half of the cases. Metastasis has never been reported [1, 3]. It shows a male preponderance with majority of cases seen during the first decade of life [1], [4], [5] as was the case in our reported observation; he is a 3-and-a-half-year-old boy. Clinically, the patient usually presents with a small, painless, slow growing soft tissue swelling which has a predilection for back and thigh. Other less common sites include the anterior chest, perineum, extremities [1], [3], [5]. In our case, the tumour was in the left shoulder. This tumour is usually superficial located in the dermis and hypodermis [6]. The histogenesis of giant cell fibroblastoma has been a topic of considerable debate amongst researchers. When examined, giant cell fibroblastoma appears as an encapsulated, gray-white, gelatinous lesion [2], [7], as in our observation. Previously, giant cell fibroblastomas were considered as a juvenile form of dermatofibrosarcoma protuberans (DFSP) but recent studies suggest that both these lesions are on a spectrum of same entity as confirmed by clinical, morphological, immunophenotypical and molecular studies [4], [6], [8], [9]. Both lesions share common clinical features of male predominance, slow growth, painlessness, and anatomical location of mainly trunk. Morphologically, both GCF and DFSP shows mainly dermal or subcutaneous location and rarely superficial skeletal muscle involvement, honeycomb and parallel growth patterns, sparing of adnexa, myxoid changes and prominent vasculature [8] Although both giant cell fibroblastoma and dermatofibrosarcoma protuberans share several clinical as well as morphological features, we still differentiate them solely on the basis of histological findings. Vimentin is the only immunohistochemical detection marker that has been found to consistently stain giant cell fibroblastoma [6], [10]. Due to its rarity and unfamiliarity, GCF was misdiagnosed in the past as sarcoma, which leads to aggressive therapy for this tumour. Shmooker et al. in their study of 28 cases of GCF revealed that 14 cases were initially diagnosed as sarcoma which led to wider surgical excision in four patients, one of whom also received radiation therapy [4]. In their report of three cases of giant cell fibroblastoma misdiagnosed two cases as sarcomas which led to more aggressive therapy [11]. The differential diagnosis can be made with childhood fibrosarcoma (FSI) and inflammatory myofibroblastic tumour (IMD) [12]. Treatment includes removal of the tumour with wide excision and these patients do not require any form of chemotherapy. Although local recurrence is common, long term outcome of this tumour is very good after excision of the tumour [13].

5. Conclusion

Giant cell fibroblastoma is a rare childhood tumour of intermediate grade malignancy which share several features with dermatofibrosarcoma protuberans. Despite having many modern diagnostic modalities at our hand, we still differentiate these two lesions solely on histological examination and all that we need is thorough histological examination of the entire tumour mass. In addition, it requires long monitoring because it’s recurrent nature.

6. References

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