Solitary nodule on the shoulder of an 11-year-old child

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Abstract

Plexiform fibrohistiocytic tumor is an uncommon soft tissue neoplasm of intermediate malignancy, most frequently occurring as a painless, slow-growing nodule that shows a distinct predilection for children and young adults. We report a healthy 11-year-old boy presenting with a 1-year history of an asymptomatic cutaneous nodule on his left shoulder. Histopathological and immunohistochemical analysis confirmed a diagnosis of plexiform fibrohistiocytic tumor. Despite following a usually benign clinical course, this neoplasm is prone to frequent local recurrence and occasional metastatic ability. It should be considered in the differential diagnosis of an enlarging nodule in pediatric patients.

Keywords: plexiform fibrohistiocytic tumor, fibrohistiocytic proliferations, soft tissue neoplasm

Introduction

Fibrohistiocytic tumors represent a vast and heterogenous group of cutaneous lesions. Plexiform fibrohistiocytic tumor (PFHT) is a rare soft tissue neoplasm of intermediate malignancy, first described by Enzinger and Zhang in 1988 [1] and distinguished by its characteristic histological pattern.

Case Synopsis

A previously healthy 11-year-old boy was referred to our department with a 1-year history of a slowly enlarging, asymptomatic cutaneous lesion on his left shoulder. Examination revealed a 10mm erythematous, well-circumscribed, smooth nodule of rubbery consistency, nontender upon palpation (**Figure 1A)**. On dermoscopy, a structureless, homogenous pink lesion was observed (**Figure 1B)**. The remaining physical examination disclosed no additional abnormalities.

An excisional biopsy was performed for both diagnostic and therapeutic purposes. Hematoxylin and eosin stain revealed a well-circumscribed dermal and hypodermal proliferation composed of histiocytic-like cells aggregated in nodules, surrounded by spindle cell fascicles in a plexiform configuration (**Figure 2**). Through immunohistochemical analysis, lesional cells stained positive for CD68 and negative for S100 and CD34 (**Figure 3**). These features were compatible with a diagnosis of PFHT. In the first specimen, the tumor extended to the deeper surgical margin. A second wider excision was performed, which showed no signs of residual neoplasia. The patient remains

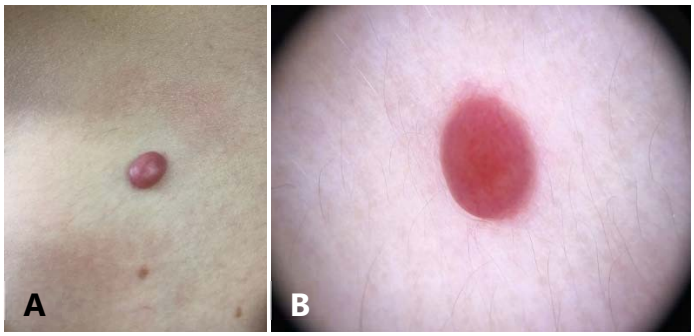

Figure 1. ***A)*** *Solitary, erythematous, smooth nodule on the patient's left shoulder.* ***B)*** *Dermoscopy showed a structureless, well-circumscribed homogenous pink lesion.*

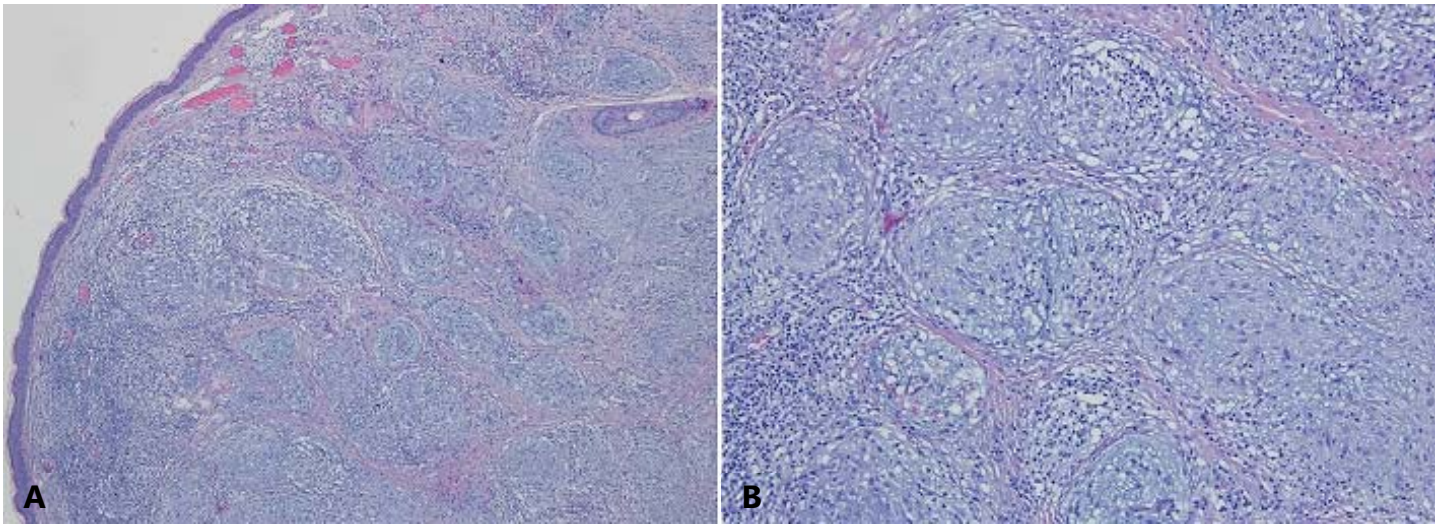

Figure 2. *Histopathological analysis revealed a dermal and hypodermal neoplasm, with histiocytic-like cells aggregated in nodules, surrounded by spindle cell fascicles in a plexiform configuration. H&E,* ***A)*** *40×.* ***B)*** *100×.*

under follow-up, with no evidence of local recurrence after 7 months of surveillance.

Case Discussion

Since the first description of this distinct neoplasm in 1988, about 200 cases have been reported in the literature. Although its age of presentation ranges from two months to 77 years [2, 3], PFHT shows a distinct predilection for children and young adults, with 70% of cases occurring in individuals under 20 years of age [3]. A congenital origin has been described in only very few patients [3-5]. Despite a female prevalence being initially suggested, recent studies demonstrate a similar distribution between genders [2-4].

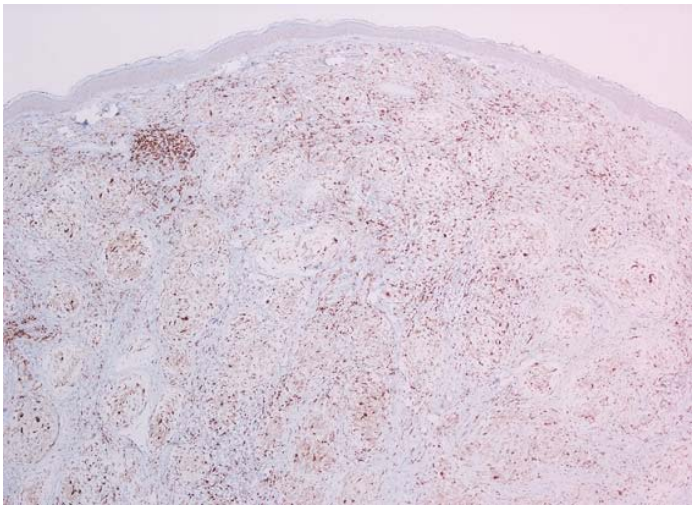

Figure 3. *Immunohistochemical staining demonstrated positivity for CD68, 40×.*

It clinically presents as a painless, solitary, slow-growing dermal or subcutaneous nodule [1-6] or, less frequently, as a firm, indurated plaque [4-6]. In rare instances, ulceration can develop [4]. Up to 50% of cases occur on the upper extremities, followed by the lower extremities, trunk, and head and neck regions [1, 5, 6]. Lesions show an indolent clinical course and can occasionally remain stable in size for several years [3].

Diagnosis relies primarily on the unique histopathological pattern of PFHT [2]. The neoplasm is typically located in the deep dermis, with possible extension to the hypodermis and other deeper subcutaneous layers [6]. It is composed of two main cell populations, arranged in a characteristic plexiform configuration. Histiocytic-like cells, admixed with multinucleated osteoclast-like giant cells, aggregate to form nodules. Fibroblast-like spindle cells, on the other hand, are organized in fascicles that interconnect adjacent nodules [1, 5]. Mild cellular pleomorphism and low mitotic activity are commonly observed. However, variants with significant cytological atypia are possible, albeit infrequently observed [1, 5, 6]. Likewise, vascular invasion has been rarely reported [1, 3]. Immunohistochemical analysis is useful in

supporting a diagnosis of PFHT. In this regard, histiocytic and osteoclast-like cells show a strong positivity for CD68, whereas spindle cells express smooth muscle actin. Additionally, PFHT cells are negative for S100, desmin, cytokeratins, and CD34 [5, 6].

The differential diagnosis is vast and must include other lesions with a plexiform growth pattern, namely fibrous hamartoma of infancy, plexiform neurofibroma, plexiform spindle cell nevus, fibromatosis (extra-abdominal desmoid), soft tissue giant cell tumor, or cellular neurothekeoma [1, 4-6].

Surgical excision is the primary reported treatment. The local recurrence rate is high and ranges from 12.5 to 50% in the published literature [4-6], reflecting the infiltrative nature of the tumor and the consequent difficulty in achieving a complete excision [6]. Regional lymph node metastases have been reported in 2.4% of cases. Despite its rarity, distant dissemination to the lungs has also been described in 1.9% of patients [2], having resulted in one fatal outcome [4]. At present, no clinical or histopathological features of PFHT can reliably correlate with its behavior and metastatic capacity [1, 5, 6]. Although safety margins have not yet been established, wide excision with negative resection margins is recommended in all patients, including children, to minimize the possibility of recurrence [1, 6].

Conclusion

In summary, PFHT is a rare neoplasm that should be considered in the differential diagnosis of cutaneous nodules in newborns and infants [1]. Regardless of its usually benign clinical course, PFHT is prone to frequent local recurrence and occasional metastatic ability. As such, prompt treatment and a thorough long-term surveillance are recommended.

Potential conflicts of interest

The authors declare no conflicts of interests.

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