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CASE REPORT

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Dermatofibrosarcoma protuberans in a child, that's why soft tissue lesions are not always innocent in children

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Abstract

Pediatric soft tissue tumors should be always operated and sent for a biopsy.

KEYWORDS

case report, children, dermatofibrosarcoma protuberans, soft tissue tumors

1 | INTRODUCTION

Soft tissue tumors are quite different in children than in adults. The incidence of malignancy is rare. Herein, we describe a case of a girl with a small soft tissue tumor in the chest wall, that although its initial diagnosis was that of a benign mass, the pathology showed dermatofibrosarcoma protuberans.

Soft tissue lesions usually represent a diagnostic dilemma since there is a large and heterogeneous group of masses in children. Although often the history and physical examination findings are sufficient for the diagnosis which most of the times is a benign mass, there are cases in which the pathology results after the surgical excision are different than the expected.[1] Such a case is the one it is described below in which the surgical excision led to early diagnosis of a dermatofibrosarcoma protuberans (DFSP), a rare fibroblastic soft tissue sarcoma of low to intermediate malignancy.[2]

2 | CASE REPORT

An 8-year-old girl presented for evaluation of a plaque-like skin lesion on her left chest wall that had slowly grown for the last 2 years. In the beginning, it was palpated only under her skin, and lately, it was obvious by visualization. The lesion was approximately 0.5 cm in maximum diameter in size. It was too small and with a light violet color, fact that did not troubled us to take a photograph of it, since it resembled in appearance with a simple granuloma. The ultrasound examination revealed an ovoid, well-demarcated lesion in the subcutaneous region with dermal involvement and without clear separation from the underlying muscle (pectoralis major). Internal echotexture of the mass was heterogeneously hypoechoic, with small echogenic foci. There was no posterior acoustic enhancement or shadow (Figure 1).

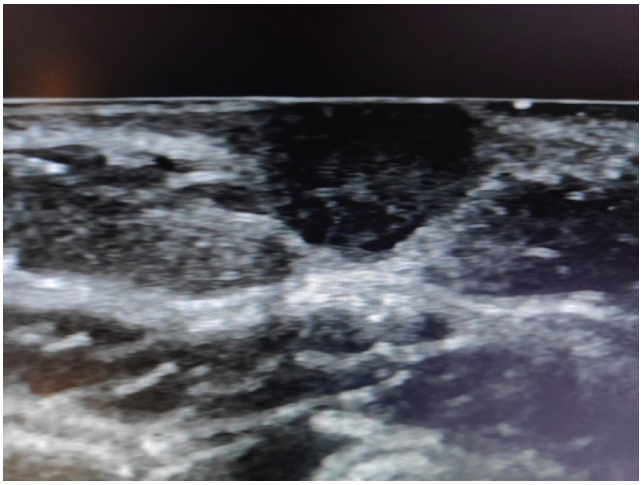
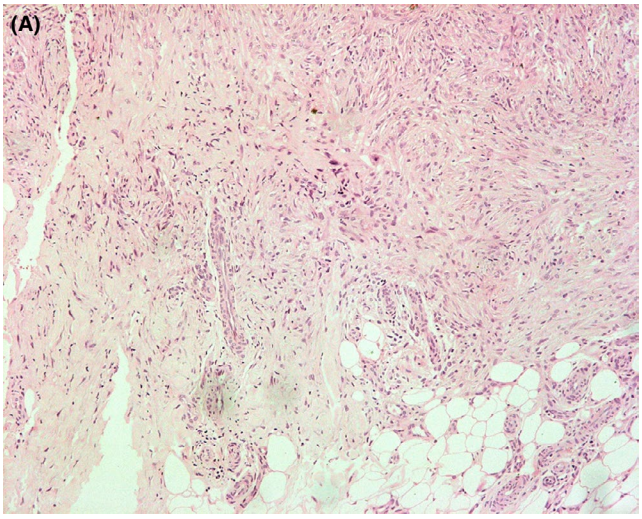


FIGURE 1 An ultrasound image in axial plane. A nodular soft tissue mass involving the skin and subcutaneous adipose tissue is seen. No calcifications or necrosis are present



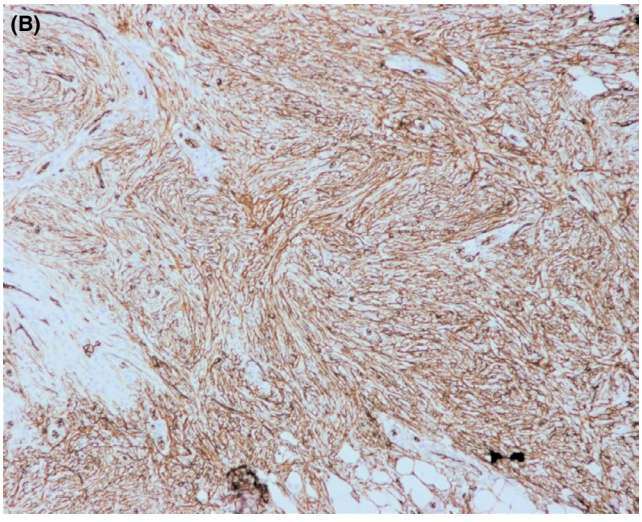


FIGURE 2 A, Spindle cells set within collagenous stroma in dermis and honeycomb appearance of subcutaneous fat (H&E x100). B, Strong immunostain for CD34 (x100)

Although this mass did not trouble us in appearance, the patient underwent surgical excision and biopsy as we do in every soft tissue lesion in our clinic. The pathology report described the specimen as having a polypoid appearance with raised epidermis macroscopically. During cut-up sectioning, a nodular lesion was observed, centered mainly within the dermis and focally in the subcutaneous tissue. It was white-gray and firm. Histologically, the epidermis was hyperplastic with hyperkeratosis and elongated rete ridges. The dermal lesion consisted of spindle cells developed in a storiform to whorled pattern. The cells had similar morphology with eosinophilic cytoplasm, monomorphic nuclei, and sparse mitotic activity. Tumor cells infiltrated fat lobules in a honeycomb pattern. The stroma was collagenous. The differential diagnosis based on hematoxylin and eosin included DFSP and Dermatofibroma. Immunohistochemically, the cells strongly and diffusely expressed CD34 and were negative for FXIIIA and S100. A confident diagnosis of DFSP was made. Moreover, no fibrosarcomatous transformation was noticed (Figure 2A,B).

After the diagnosis, the patient underwent extensive imaging and hematological testing which were without pathological findings. Although the margins of the removed mass were healthy, we decided extended surgical excision (2-cm boundaries from the previous postoperative scar) to decrease the possibility of recurrence. (Figure 3) The child's postoperative course was uneventful, and she will be examined every 6 months for the next 3 years for palpation of her scar tissue and examination of her regional lymph nodes to exclude recurrence or the possibility of metastases.

3 | DISCUSSION

A wide spectrum of entities may give rise to soft tissue masses in children, including both neoplastic and nonneoplastic lesions. Firm plaques and nodules appear many times as skin lesions in the pediatric population and worry the clinician with the broad-spectrum differential diagnosis. The decision of biopsy most of the time will be driven by the evolution and

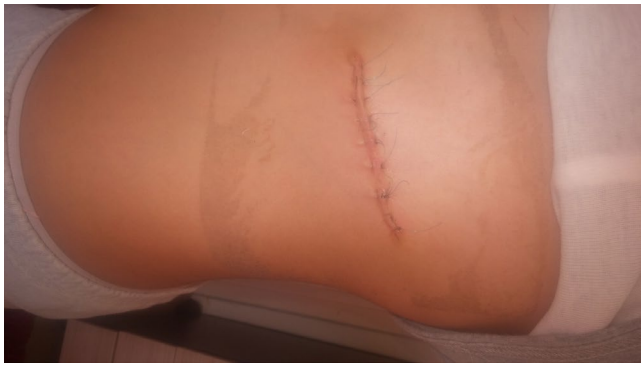


FIGURE 3 Trauma after extended surgical excision

location of the tumor, but sometimes the absence of characteristic features can delay this decision.[3]

Dermatofibrosarcoma protuberans belongs to the category of tumors in which the diagnosis often delays because of either general hesitancy to biopsy or the clinical appearance that rarely raises the suspicion of a neoplastic tumor.[3] In our case, although the parents observed the lesion in the chest wall of their girl a long time ago, they considered it as an innocent mass and were concerned only after the increase in its size.

Dermatofibrosarcoma protuberans was originally described as a distinct clinicopathologic entity by Darier and Ferrand in 1924.[4] Term "dermatofibrosarcoma protuberans" was proposed and coined by Hoffman in 1925, but its occurrence in childhood was not mentioned in the literature prior to 1957.[4,5] Most of the times the mass affects the trunk, accounting for almost half of all cases and rarely other parts of the body.[6] It typically appears as an asymptomatic plaque with a hard consistency fixed to the skin with no deep layers and a size that ranges between 1 and 5 cm and increases over the years as in our case. Mainly, the diagnosis is made in adulthood although the mass grows slowly with lack of symptoms since childhood. In the most recent review study, it is referred that only six children under 14 years old had DSFP out of 159 people (3.7%), although in the literature, the proportion in the pediatric population is between 6% and 20%.[4]

The radiologic appearance of DFSP is not pathognomonic. The typical ultrasound appearance is that of a nodular soft tissue mass involving the skin and subcutaneous adipose tissue without calcifications. However, when we have to deal with a superficial, nodular, or multinodular soft tissue mass, which involves the skin and subcutaneous adipose tissue, the DFSP must be at the forefront of our differential diagnosis.[7]

The histology of DFSP with storiform or whorled pattern and honeycomb appearance of subcutaneous fat is considered the gold standard features for the diagnosis. Multiple variants, not rarely seen in a busy dermatopathology laboratory, can raise difficulties in the differential diagnosis. Giant cells, melanin pigmentation (Bednar tumor), pseudocystic changes, myxoid stroma, and sarcomatous transformation are all well-described variants, potentially causing difficulties as regards the final diagnosis.[8,9] Moreover, loss of CD34 expression in sarcomatous changes can lead to misdiagnosis of this entity. Immunohistochemically, the tumor expresses CD34 and vimentin, while it is negative for FXIIIA, S100, Desmin, SMA, AE1/AE3, MelanA, and HMB45. The most common fusion is COL1A1-PDGFB.[10] Histologically, the differential diagnosis includes dermatofibroma (positive for FXIIIA and negative for CD34), melanoma and melanocytic tumors (S100, MelanA, and HMB45 positive), and leiomyoma/leiomyosarcoma (Desmin and SMA positive).

The treatment of choice in nonmetastatic DSFP is the complete surgical resection with wide margins. Primary surgical closure is made especially in redundant soft tissues such as chest, abdomen, and back. Sometimes, skin grafting or flap reconstruction is essential.[4] The wide margins in resection are important because of the recurrence rate which ranges from 26% to 60% in cases of conservative resection in contrast to wide local excision with 2-3 cm margins from the tumor boundaries with much lower recurrence rates (0%-30%).[11] In children, the recurrence rate is approximately 9%; however, very little data exist on relapsed pediatric DFSP.[2] In our case, although the surgical resection was in healthy margins after the pathology report, we decided to remove extended tissue to reduce the recurrence rate. Moreover, we adopt a close follow-up examination period according to the literature during the first 3 years every 6 months followed by an annual follow-up because 50% of recurrences develop in the first 3 years after resection.[4]

CONCLUSION

In conclusion, clinicians must adopt a high index of suspicion in pediatric soft tissue masses and prefer excision more easily because although these masses resemble benign lesions, only histological sampling is the key for the right diagnosis and early treatment.

CONFLICT OF INTEREST

The authors have no conflicts of interest.

AUTHOR CONTRIBUTIONS

KK and SD: designed, reviewed, and revised the manuscript. SF: drafted the initial manuscript and contributed to the interpretation of data. CD: drafted the initial manuscript and contributed to the acquisition of data. MA: drafted and designed the initial manuscript. AG and PC: drafted and designed the initial manuscript. All the authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

ETHICS APPROVAL AND CONSENT

Signed consent was obtained from the patient's mother for this case report as the patient herself was unable to consent. The patient's mother was also made aware that the information taken was used for publication.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

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