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Pathology

Pediatric Nodular Fasciitis of the Back: Imaging Pitfalls and Pathological Certainty

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Abstract | **Case Report**

Background: Nodular fasciitis is a benign, self-limiting myofibroblastic proliferation that is often clinico-pathologically mistaken for a malignant soft tissue sarcoma due to its rapid growth and high cellularity. While it can occur anywhere, it is frequently found in the upper extremities and trunk of young adults. ***Case Presentation:*** We report the case of a 9-year-old patient presenting with a rapidly enlarging, subcutaneous mass located on the back. Initial clinical and radiological assessments were suggestive of a schwannoma or lipoma. A complete surgical excision was performed and the diagnosis of nodular fasciitis was confirmed finally through histological and immunohistochemical examination. ***Conclusion:*** Nodular fasciitis remains a "great imitator" in soft tissue pathology. This case highlights the essential role of immunohistochemistry in preventing diagnostic pitfalls and avoiding overly aggressive surgical interventions for a benign lesion with an excellent prognosis.

Keywords: Nodular fasciitis, Myofibroblastic proliferation, Pediatrics, Ultrasonography, Immunohistochemistry, Differential diagnosis.

INTRODUCTION

Nodular fasciitis (NF) is a benign, self-limiting myofibroblastic proliferation that typically arises from the superficial fascia [1]. Historically characterized as a "pseudosarcomatous" lesion, NF is notorious for its rapid clinical growth and high cellularity, features that can frequently lead to a misdiagnosis of mesenchymal malignancy [1,2]. In the pediatric population, where soft tissue tumors encompass a broad spectrum of benign and aggressive entities, achieving an accurate preoperative diagnosis is paramount to guide conservative management [3,4].

The clinical utility of imaging in the diagnosis of NF remains limited. Ultrasonography, while often the first-line diagnostic modality due to its accessibility and non-ionizing nature in children, typically reveals non-specific features [5]. On grayscale imaging, NF commonly presents as a well-circumscribed, hypoechoic subcutaneous mass with variable internal vascularity [5]. Such findings often overlap with those of other benign neurogenic or adipocytic lesions [5,6]. Specifically, the proximity of these nodules to fascial planes or peripheral nerves frequently leads to a radiological suspicion of schwannoma, while their subcutaneous location may prompt a diagnosis of an atypical or deep-seated lipoma [7,8].

Given these diagnostic pitfalls in imaging, immunohistochemistry (IHC) has emerged as the definitive cornerstone for establishing a correct diagnosis. While the "tissue-culture" morphology of spindle cells is suggestive, IHC allows for the critical exclusion of neural (S100, SOX10) and myogenic (desmin) markers, while confirming a myofibroblastic lineage through the characteristic expression of smooth muscle actin (SMA) [6].

This report presents a case of nodular fasciitis of the back in a 9-year-old patient. By examining the discrepancy between initial ultrasonographic suspicions—which suggested a schwannoma or lipoma—and the final pathological findings, we aim to emphasize the indispensable role of immunohistochemistry in the differential diagnosis of pediatric soft tissue masses.

CASE REPORT

A 9-year-old male with an unremarkable past medical history presented with a subcutaneous swelling on his back. The patient's parents reported that the mass had appeared recently and was rapidly enlarging. Clinical examination revealed a firm, non-tender, and mobile subcutaneous nodule located in the dorsal region, measuring 3 cm in its greatest dimension. There were no overlying skin changes or associated neurological symptoms.

Initial assessment was performed via soft tissue ultrasonography, which identified a well-defined, heterogeneous mass situated within the subcutaneous tissue. The lesion appeared predominantly hypoechoic with internal echogenic areas. Notably, color Doppler imaging demonstrated significant internal vascularity. Given the patient's age, the anatomical site, and the sonographic appearance, the initial diagnostic hypotheses included a schwannoma or a lipoma with atypical features.

Due to the rapid growth and the vascular nature of the lesion on imaging, an excisional biopsy was performed. Macroscopic examination of the specimen showed a non-encapsulated, solid, grayish-white nodular mass with a slightly myxoid and firm consistency.

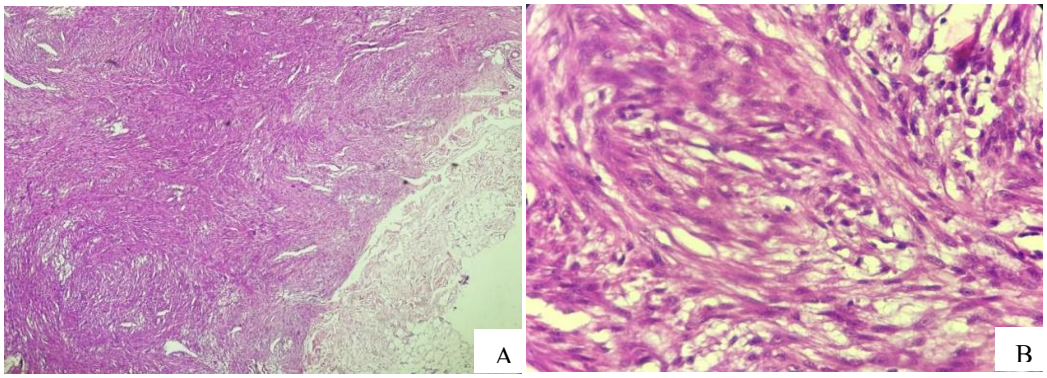

Figure 1: Nodular fasciitis: (A) Low-power view showing a non-encapsulated, spindle cell proliferation (4x); (B) High-power view highlighting the "tissue culture-like" appearance (40x)

Microscopic analysis of the excised mass revealed a non-encapsulated but well-circumscribed proliferation of spindle-shaped cells. These cells, identified as myofibroblasts, were arranged in short, irregular bundles and fascicles, creating a characteristic "tissue-culture" appearance. The cells exhibited plump, ovoid nuclei with fine chromatin and small, inconspicuous nucleoli, without significant nuclear pleomorphism or atypia.

Mitotic figures were readily identified, reflecting the rapid clinical growth of the lesion; however, no atypical mitotic forms were observed (Figure 1).

To further characterize the lesion and definitively differentiate it from the initial radiological suspicions of schwannoma or lipoma, an immunohistochemical (IHC) panel was performed :

Smooth Muscle Actin (SMA) Demonstrated strong and diffuse cytoplasmic staining in the spindle cells, displaying a characteristic "tram-track" pattern. This finding confirmed the myofibroblastic nature of the proliferation.

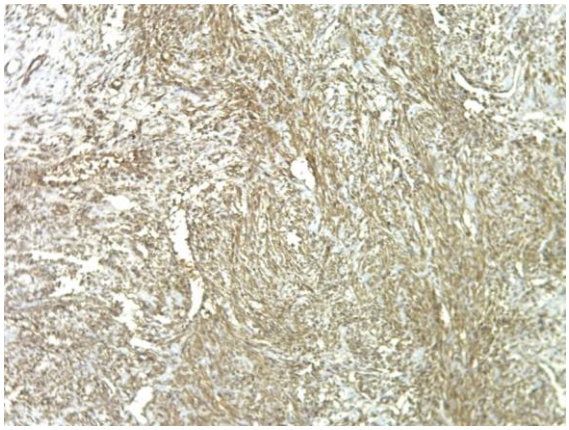

Figure 2: Immunohistochemical staining showing strong and diffuse cytoplasmic expression of smooth muscle actin (SMA) within the spindle cell proliferation

SOX10 was consistently negative within the spindle cell population, effectively excluding a schwannoma or other peripheral nerve sheath tumors. Desmin was Negative ruling out a primary myogenic tumor such as rhabdomyosarcoma. CD34 was Negative, which helped exclude a solitary fibrous tumor or dermatofibrosarcoma protuberans (DFSP). Caldesmon was Negative, further distinguishing the lesion from a true smooth muscle tumor (leiomyoma). The combination of the classic "tissue-culture" morphology and the specific IHC profile (SMA+/S100-) led to the definitive diagnosis of nodular fasciitis.

DISCUSSION

Nodular fasciitis (NF) remains one of the most challenging benign lesions in soft tissue pathology due to its ability to mimic malignant processes both clinically and morphologically [2]. In the pediatric population, while NF is well-documented, its presentation can frequently lead to diagnostic uncertainty, especially when initial imaging points toward more common localized tumors such as schwannomas or lipomas [9].

In this case, the ultrasonographic findings of a heterogeneous, vascularized mass were instrumental in the initial suspicion of a schwannoma [5]. Schwannomas in children, though rare, often present as well-circumscribed, hypoechoic, and vascularized nodules on Doppler. Similarly, while lipomas are typically isoechoic to fat, atypical or spindle-cell variants can present with increased heterogeneity and vascularity, leading to preoperative confusion [10]. The rapid growth reported in this 9-year-old patient—3 cm in just three weeks—is a classic clinical hallmark of NF but is often misinterpreted by clinicians as a sign of malignancy or aggressive neurogenic growth.

The definitive diagnosis of NF relie on the recognition of the "tissue-culture" growth pattern, but immunohistochemistry (IHC) serves as the "judge of peace" to exclude mimics [6].

The consistent negativity for S100 and SOX10 in our case was the most critical step. In a 9-year-old, a nerve sheath tumor would show diffuse and intense staining for these markers, which was entirely absent here.

The diffuse expression of Smooth Muscle Actin (SMA) confirmed the myofibroblastic lineage. It is important to note that the staining in NF is typically focal or peripheral within the cell (the "tram-track" appearance), unlike the dense, bundles-like staining seen in true leiomyomas.

Although the mitotic index was high, the lack of cytological atypia and the absence of Desmin staining effectively ruled out pediatric rhabdomyosarcoma or other high-grade spindle cell sarcomas.

While IHC is often sufficient, it is now understood that over 90% of NF cases harbor a specific t(17;22) translocation, leading to the MYH9-USP6 gene fusion [11]. This molecular signature has redefined NF from a reactive inflammatory process to a "transient neoplasm"[12,13]. In complex cases where IHC might be equivocal, the detection of USP6 rearrangement via FISH or NGS provides a definitive "gold standard" for diagnosis, preventing unnecessary radical surgeries in children.

CONCLUSION

This case highlights that in the presence of a rapidly growing subcutaneous mass in a child, nodular fasciitis should always be considered, even when imaging suggests a neurogenic or lipomatous origin. A meticulous pathological examination supported by a targeted IHC panel (SMA+/S100-) remains essential to ensure a benign diagnosis and avoid over-treatment.

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