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Case Report

Nodular fasciitis-often a tumor mimic

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ABSTRACT

Nodular fasciitis is a benign proliferation of fibroblasts and myofibroblasts. It is frequently mistaken for a sarcomatous lesion because of its rapid growth, imaging features and aggressive pathological findings. Though imaging plays a vital role in evaluating these lesions, strong suspicion with a careful histopathology is the final answer. We present a patient with nodular fasciitis involving the upper extremity. A 15-year-old boy presented with complaints of progressively increasing right wrist swelling since 2 months, and had been having severe pain on exertion. On clinical examination the swelling was tender. MRI revealed T2 hyperintense lesion along the volar-lateral aspect of wrist, completely encasing the abductor pollicis longus tendon and indenting the extensor pollicis brevis tendon. Per operatively, the tumor was seen to involve the abductor pollicis longus tendon sheath and insinuating between the tendons. Excision biopsy was performed and histopathology confirmed the diagnosis of nodular fasciitis.

Keywords: Benign, Fibroblast, Myofibroblasts, Tender

INTRODUCTION

Nodular fasciitis is a benign rapid proliferation of fibroblast and myofibroblasts which often mimics a malignant lesion. It usually affects subcutaneous tissues, muscles, tendons and fascia. It was first described by Konwaler in 1955 as a subcutaneous pseudosarcomatous fibromatosis.[1] Nodular fasciitis is a relatively uncommon entity in children and adolescence. The peculiarities of this lesion which makes it unique are its self-limiting potential, spontaneous regression, often being misdiagnosed as malignant tumor and less recurrence rate. The lesion commonly occurs in upper extremity and chest; however, hand is a relatively uncommon site of occurrence in upper extremity.[2-4] Our patient is a 15 years old boy presented with progressively increasing painful swelling in the wrist. Though there are literatures on nodular fasciitis, it is important to create more awareness about this pathology as limited resection of the lesion is curative in the majority.

CASE REPORT

A 15-year-old boy presented with complaints of right wrist swelling since 2 months, progressively increasing in size. The patient complained of severe pain on exertion. There were no systemic symptoms like fever, loss of appetite or weight loss. There was no history of any other joint pain/ stiffness. The patient denied history of trauma.

On clinical examination, the swelling was tender. There was no redness or pulsation in the swelling. Ultrasound showed a well-defined hypoechoic lesion in the volar aspect of the wrist with internal vascularity and we suggested further evaluation with MRI. MRI revealed a well-defined T1W hypointense lesion appearing hyperintense on PDW sequences along the volar-lateral aspect of the wrist, almost completely encasing the abductor pollicis longus tendon (Figure 1). The lesion was seen indenting the extensor pollicis brevis tendon.

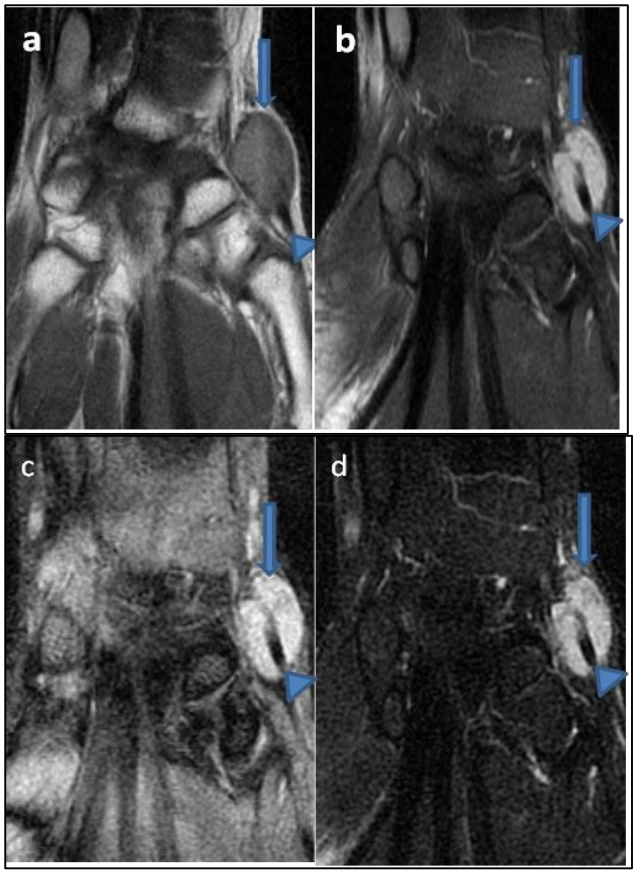

Figure 1 (A-D): Coronal T1W image (a) show a well-defined hypointense lesion (arrow) and PDW Fat saturated image (b) show hyperintense lesion encasing the abductor pollicis longus tendon (arrowheads). Coronal GRE (c) and STIR images (d) show the hyperintense lesion (arrows) almost completely encasing abductor pollicis longus tendon (arrowheads).

The lesion was hyperintense on short tau inversion recovery, STIR, sequence and showed high signal intensity on gradient echo, GRE, sequence (Figure 1 C and D). There was no evidence to suggest phleboliths or flow voids within the lesion which would favor a vascular malformation. Axial images show the tendon encasement better (Figure 2).

Differential diagnosis of giant cell tumor of the tendon sheath, soft tissue sarcoma, fibromatosis and nodular fasciitis were considered based on imaging.

Per operatively, the tumor was seen to involve the abductor pollicis longus tendon sheath and insinuating between the tendons. There was no vascular invasion. No abnormal lymphnodal disease identified during the surgery. Excision biopsy was performed.

Histopathology confirmed the diagnosis of nodular fasciitis. The patient's parents were educated about the disease and appropriate follow up was advised.

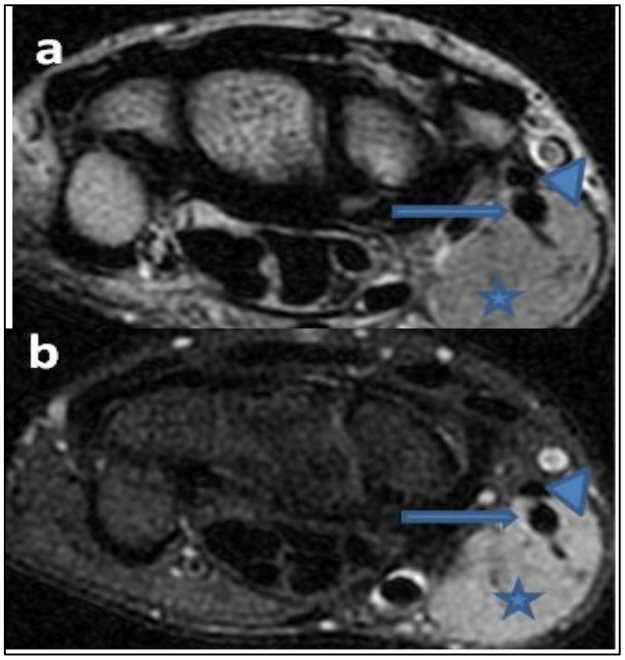

Figure 2 (A and B): Axial T2W FSE and PD FS images show the well-defined hyperintense lesion (asterisk) almost completely encasing.

DISCUSSION

Nodular fasciitis, a benign rapid proliferation of fibroblasts and myofibroblasts, occur commonly in young adults, age ranging between 20 to 40 years.[5] The incidence in children is around 10%.[5,6] This rare entity was described using various terms earlier like nodular fibrositis and subcutaneous fibromatosis. Later the term nodular fasciitis was used by Price et al as it presented like a tumorous nodule and involved subcutaneous fascia/ fat extending sometimes to involve deep fascia and muscles.[7] According to 2013 WHO classification, this is considered as benign lesion of fibroblastic/ myofibroblastic tumor class.[8] Though the pathogenesis is unclear, recent studies have elucidated the role of gene rearrangement of ubiquitin specific protease as a specific finding in these cases.[9]

Clinically, nodular fasciitis present as solitary rapidly enlarging subcutaneous painful swelling. It becomes clinically apparent in few weeks and usually measures around 2 to 3 cm.[10] The most common location is arm/forearm followed by trunk and lower extremity.[10] These lesions are mistaken for malignant lesions like sarcoma due to their rapid growth, increased mitotic activity, and infiltrative tendencies, leading unnecessarily to extensive surgical resections.[11]

Wu et al evaluated the role of MRI in differentiating nodular fasciitis and other soft tissue lesions, and came with four different helpful signs on MRI. The most frequent sign which they found helpful to identify nodular fasciitis was "fascia tail sign" which presented as a broad fascial base with a linear extension of the nearby

fascia.[11] Most nodular fasciitis lesions exhibited homogenous to isointense signals on T1WI and hyperintensity on T2WI in earlier studies.[12,13] On post contrast imaging, homogenously enhancing lesions may be more cellular tissue, while lesions with mild enhancement may show more myxoid changes.[11,14] Our patient had a homogenously enhancing lesion suggesting increased cellularity and was later proven histologically.

While spontaneous regression is one of its unique features, we would recommend a surgical resection for these lesions as this can often be a tumor mimic and cause diagnostic dilemma. Recurrence is rare after limited surgical resection; hence the aim should be to avoid aggressive surgery.[15]

CONCLUSION

Nodular fasciitis, an uncommon entity, needs reiteration as it can often mimic a soft tissue sarcoma both on imaging and on histopathology. A strong suspicion and careful histopathology can help us in diagnosing this self-limiting lesion. Limited resection should be the norm if nodular fasciitis is suspected and pathology should lead the way.

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