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Case Report and Review of the Literature

Posttraumatic Intramuscular Nodular Fasciitis in a 7-Year-Old Boy: Case Report and Review of the Literature

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ARTICLE INFO

Article history:
Received: 20 May, 2021
Accepted: 3 June, 2021
Published: 18 June, 2021

Keywords:
Nodular fasciitis
nodular pseudosarcomatous fasciitis
pseudosarcomatous fasciitis
subcutaneous pseudosarcomatous fibromatosis

ABSTRACT

Nodular fasciitis is a rare disease, and its diagnosis is difficult. We present a case report of a seven-year-old child with progredient swelling of the left pectoralis muscle three weeks after trauma. After histopathological diagnosis, we performed complete resection. Normally, a conservative approach with regular follow-up is regarded as appropriate since nodular fasciitis does have the capability to regress spontaneously. Since recent publications indicate the possibility of malignant transformation, the complete primary resection also has to be discussed as therapy of choice.

Case Report

A seven-year-old boy was presented to us by his parents in our emergency department. The parents reported swelling of the young boy's left shoulder/pectoralis muscle, which he had noticed after having fallen on it three weeks earlier but which he had not mentioned to them. Since the lump was becoming increasingly painful and the range of motion had reduced due to the pain, he told his parents about it after all and they then presented for consultation. Physical examination revealed a marked and pain-related reduction in range of motion of the left shoulder and arm, especially during abduction and elevation, as well as an obvious lump in the left pectoral area covered by stretched skin. Sonography was performed that showed a lesion of at least, 2.6 cm suggesting an organized haematoma. However, due to the size of the surrounding reactions, it was not entirely visible in sonography (Figure 1).

The parents were told to wait and see how the lesion develops and return for further consultation in case of persistence or progression of either the swelling or the symptoms. Physical examination two weeks later showed a progression in size leading to MR imaging of the boy's thorax. MRI revealed a round, intrapectoral mass of about 2.6 cm in size with marked perifocal edema and inhomogeneous enhancement of contrast media (Figures 2 & 3). Since this morphologic description rather matched a tumorous mass than either an inflammatory lesion or haematoma, incisional biopsy was recommended. The patient undergone this procedure two days later. The histological work-up presented the diagnosis of nodular fasciitis of the major pectoralis muscle. The complete resection with preservation of the muscle was performed one week later. The young patient presented to clinical and sonographic follow-up two months after the operation without symptoms or recurrent disease.

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http://dx.doi.org/10.31487/j.SCR.2021.06.06

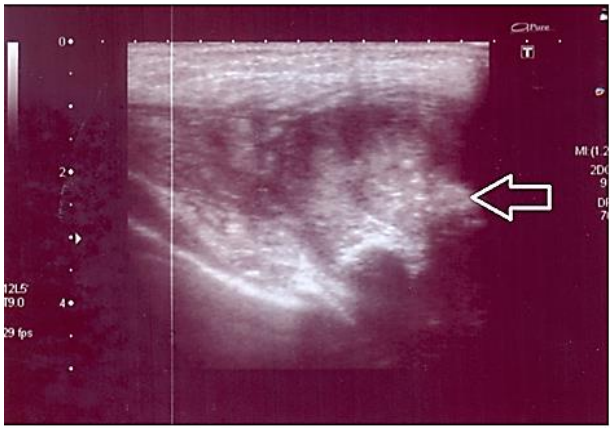
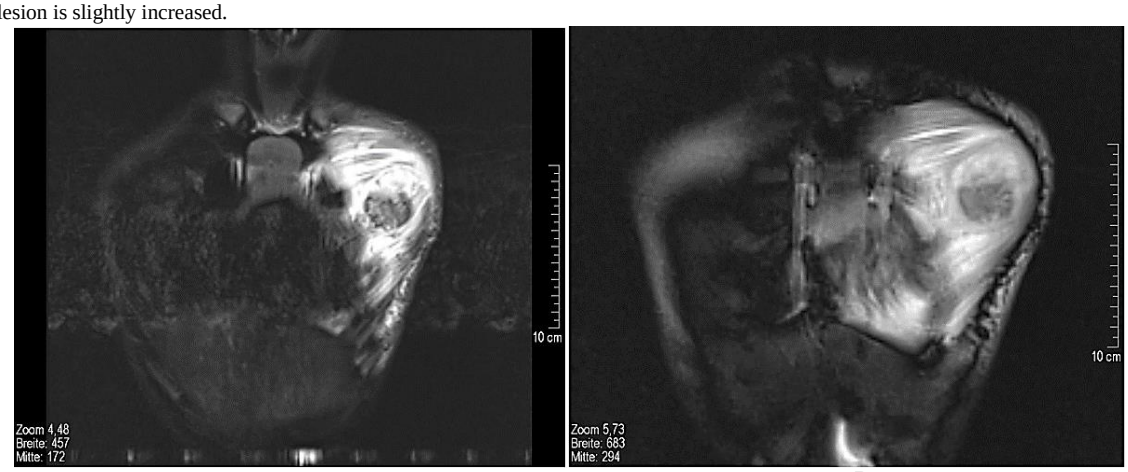
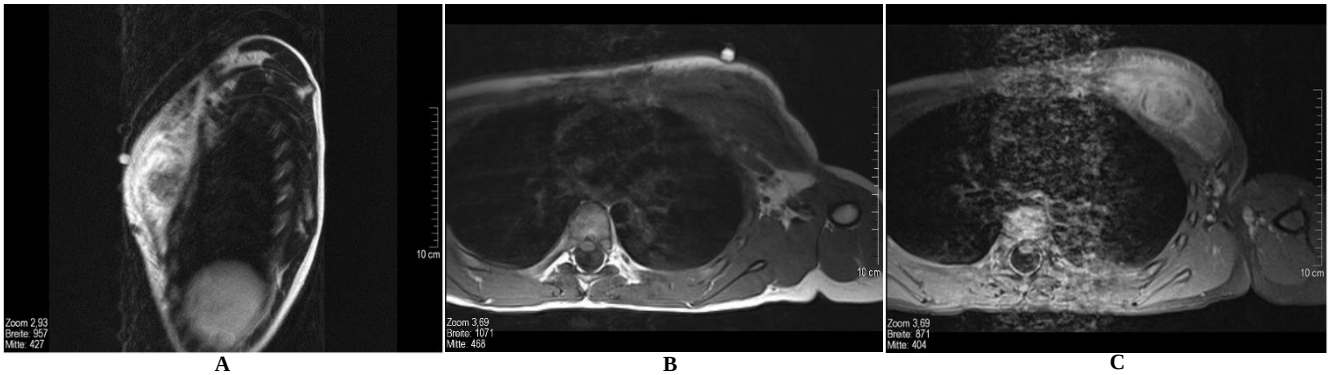


Figure 1: Soft tissue sonography of the left major pectoralis muscle: The linear probe cannot completely depict the extent of the lesion due to its size, making it necessary to use the curved probe with an imaging depth of 9 cm. An intramuscular lesion with a diameter of 2.6 cm, adjacent to the fascia and well distinguishable from other tissues, can be seen. It appears partly hyperechoic and partly hypoechoic compared to the surrounding muscle. The perfusion within the lesion is slightly increased.



A **B**

Figure 2: Magnetic resonance tomography of the thorax, without contrast media. **A)** Coronal STIR, **B)** coronal True-FISP-Sequence: marked edema of the left major pectoralis muscle. Intramuscular mass, approx. 2.6 cm diameter, which is partly isointense and partly hyperintense compared to the surrounding muscles.



A **B** **C**

Figure 3: Magnetic resonance tomography of the thorax. **A)** Sagittal T1 SE (with marking), **B)** transversal T1 TSE, **C)** transversal T1 fs after administration of contrast media. Extensive, space-occupying edema of the left major pectoralis muscle. Intramuscular mass, approx. 2.6 cm diameter, which appears partly isointense and partly hyperintense compared to the surrounding muscles in the STIR sequences and isointense in the T1w sequences. Inhomogeneous enhancement of the mass and reactive absorption of contrast media by the edema is seen after administration of contrast media.

Discussion

Nodular fasciitis (synonyms: nodular pseudosarcomatous fasciitis, pseudosarcomatous fasciitis or subcutaneous pseudosarcomatous fibromatosis) is a pseudotumorous, benign neoplasm of fibrous tissue, which is mainly found subcutaneously but can also be found in intramuscular or fascial positions of extremities or the trunk [1]. The morphologic spectrum is broad, including the classic pattern of delicate fibroblasts suspended in a myxoid matrix, granulation tissue-like areas, solid and whorled myofibroblastic proliferations with multinucleated

cells, mucoid cysts, and so-called "ancient" forms with dense, refractile strands of keloid-like collagen [2]. Most cases of nodular fasciitis occur in young adults aged 20 to 40. There is evidence that in general, both sexes are equally affected, while this question is not clearly answered for a pediatric subpopulation probably due to only very small case numbers: some authors state that more girls are affected while others found more boys to be affected [3-5]. According to Wu *et al.*, the upper extremity is the most common localization, while in a pediatric population – which is about 10% of all cases -it seems to occur predominantly on the trunk or head and neck [4, 6-8]. A variety of locations such as head, neck, parapharyngeal space, cheek, jaw, nose, eye/orbital region, tongue, parotid region, ear, knee, hand, chest, abdominal wall has been described in literature for a pediatric population [9-38].

There is only a little literature on intramuscular nodular fasciitis, such as in our case. The few existing reports state that this variant accounts for up to 10% of all findings in nodular fasciitis [1]. Not much is known on the pathogenesis of the disease. Assumptions have been made that there is an association to trauma (as in our case) [39]. The lesions can be detected easily in imaging. With ultrasound, they appear as well as distinguishable, mixed hyperechoic and hypoechoic lumps or masses (Figure 1) with slightly increased perfusion of the deep subcutaneous tissue adjacent to the fascia [39]. MRI cannot distinguish the various subtypes of nodular fasciitis such as myxoid, cellular or fibrous. Unfortunately, the morphology is very heterogenic and can mislead to the diagnosis of soft tissue sarcoma [40, 41]. When viewed in T1w sequences, lesions which are rich in cells are nearly isointense compared to musculature and hyperintense compared to fatty tissue in T2w sequences. Fibrous lesions however are hypointense in all sequences. Usually, the enhancement of contrast media is diffuse but can sometimes be located only peripherally (Figures 2 & 3) [42]. Biopsy is needed to confirm diagnosis in nearly every case. Genomic rearrangements of the USP6 locus are found in 92% (44 of 48) of nodular fasciitis [43]. Tomassen *et al.* recommended molecular analysis of untypical cases to avoid overtreatment due to misdiagnosis, which was previously described [3, 37].

In the past, nodular fasciitis was regarded as benign neoplasia and a conservative approach with follow ups and a surgical excision only in case of a progression are discussed as there are cases with a spontaneous regression and also involution after injection of steroids [44]. Newer publications indicate the possibility of a malignant transformation in isolated cases, so the primary complete surgical excision has to be discussed as therapy of choice [45, 46]. Recurrent disease is rare [8]. The differential diagnosis should include the following (depending on the lesion's localization): M. Dupuytren, desmoid tumors, neurofibromas, fibrous histiocytomas, soft tissue sarcomas and, in cases of intramuscular findings, early-stage myositis ossificans.

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