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CASE REPORT

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A Rare Case of Aggressive Fibromatosis Infiltrating Dorsal Muscles in a 6-Year-Old Patient – CT, MRI and Elastography Evaluation

Authors' Contribution:
A Study Design
B Data Collection
C Statistical Analysis
D Data Interpretation
E Manuscript Preparation
F Literature Search
G Funds Collection

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Summary

Background: The term fibromatosis or desmoid tumor refers to a group of benign fibrous growths without metastatic potential but with a significant risk of local recurrence. These lesions typically present infiltrative growth pattern with local invasion of adjacent tissues. This tendency is the reason for a relatively high rate of local recurrence, even after surgical removal. Fibromatosis is a very rare condition in general population but occurs more frequently in one of the familial cancer predispositions known as familial adenomatous polyposis (FAP) or Gardner syndrome. There are two main groups of fibromatosis: superficial (small, slow-growing lesions) and deep, also known as aggressive fibromatosis (large, rapid-growing lesions).

Case Report: We report a case of a 6-year-old boy suffering from an aggressive form of fibromatosis. The patient developed a large pathological mass extending from the neck to the loins. After incisional biopsy and histpoathological examination of the sample, a diagnosis of aggressive fibromatosis was established. During the whole diagnostic process, different imaging techniques including CT, MRI and sonoelastography were used. As the surgical treatment was not possible, the patient was finally qualified for chemotherapy.

Conclusions: Eventual diagnosis of aggressive fibromatosis is based on histopathological examination. However, it is an important condition that should be included in differential diagnosis of soft-tissue masses found in diagnostic imaging. Radiologists should be careful especially in defining the margins of infiltration in case of potential surgical treatment.

MeSH Keywords: Diagnostic Imaging • Elasticity Imaging Techniques • Fibromatosis, Aggressive

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Background

Fibromatosis or desmoid tumor is a non-malignant mesenchymal tumor which belongs to a broad group of soft tissue lesions, characterized by fibroblastic proliferation of connective tissue arising from the muscles, fascia, or aponeuroses [1,2]. Biological behavior of fibromatosis falls between benign fibromas and malignant fibrosarcomas. Although it is devoid of metastatic potential, it typically presents infiltrative growth pattern with local invasion of surrounding tissues. Local recurrence after surgical resection is common, most often within two years [3]. Fibromatosis is a rare disorder with an incidence assessed to be between two and four cases per one million a year. It represents only 0.03% of all neoplasms [4]. Desmoid tumor may develop at any age but it is more common between the age of 25 and 40, with a tendency for female predominance. However, in extraabdominal tumors, sexual predilection is

Figure 1. Thorax CT, mediastinal window, axial plane. Thickening and hypodense infiltration of right dorsal muscles. Notice small pleomorphic calcification.

not as high as in intraabdominal forms [3,5]. In most cases aggressive fibromatosis occurs sporadically but association with familial adenomatous polyposis and Gardner syndrome has been proved [6]. Two major groups of fibromatosis can be distinguished: superficial and deep. The superficial (fascial) group contains small and slow-growing lesions including e.g. palmar fibromatosis (Dupuytren disease), plantar fibromatosis (Ledderhose disease). The deep (musculoaponeurotic) forms are also called 'aggressive fibromatosis' because of rapid growth, often large size, and tendency to local recurrence after treatment. In this group, muscles of the trunk and limbs are predominantly affected [6,7].

Case Report

This article reports a case of aggressive fibromatosis presented as a large pathological mass extending from the neck to the loins in a young male patient and describes its imaging findings.

A 6-year-old boy was admitted to the Pediatric Oncology, Hematology and Transplantology Clinic in Wrocław for further diagnosis of the thoracic wall tumor. History taking revealed that the child developed non-tender and firm in consistency swelling located below the right scapular angle. The lesion was noticed by his mother about two months earlier and it gradually enlarged during that time. Blood test results were within normal limits, apart from the CRP level, which was slightly elevated. On physical examination, a fixed, firm mass was palpable, not only below the right scapula, but also on the neck and in the right lumbar region. The range of movement in the patient's right arm was limited. As his clinical presentation was highly suggestive of malignancy, incisional biopsy from a visible mass was performed. Because of a high suspicion of malignant tumor, large extent of the lesion, and necessity of excluding the metastases, especially in lungs, computed tomography scanning was decided to be carry out as the first diagnostic imaging method.

CT examination of the neck, thorax, abdomen and pelvis was performed, using a 128-slice Siemens Somatom computed tomography scanner before and after i.v. contrast medium

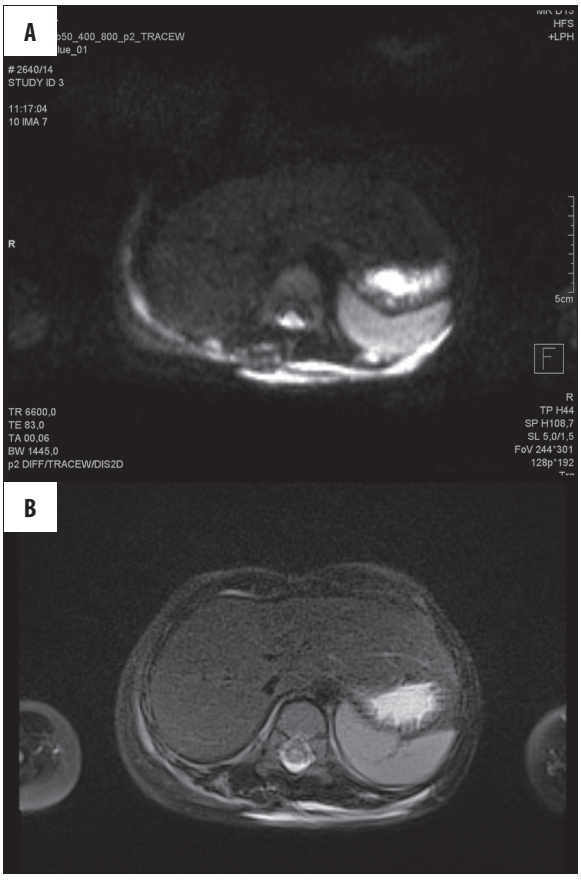

Figure 2. Abdominal MRI. DWI, b-value 50 (**A**) and BLADE sequence, T2W, fat saturation (**B**). Infiltration is visible on both sides in the dorsal region. Notice that the margins of hyperintense infiltration reache farther in peripheral direction on DWI images than on BLADE images.

injection. CT scans revealed a pathological diffuse soft tissue mass located between muscles in the posterolateral aspect of the right part of the body, extending from the neck through the thoracic wall to the right lumbar region. The bulk of the mass was of heterogeneous density with a few pleomorphic calcifications and demonstrated a little enhancement after intravenous contrast medium injection. Poorly defined margins suggested infiltration of the surrounding muscles which made a diagnosis of malignancy more feasible (Figure 1). Despite the fact that the mass abutted the right scapula, there was no visible evidence of bone destruction.

CT scans did not reveal any pathology of thoracic or abdominal and pelvic organs. However, a few slightly enlarged mesenteric lymph nodes were visible.

Based on histopathological examination, the mass was confirmed as fibromatosis. Considering large extent and invasive growth pattern, a final diagnosis of aggressive fibromatosis was established.

For more precise assessment of the infiltration, MRI examination was carried out using a Siemens Avanto 1,5T MRI scanner. On MR images the lesion appeared to be of iso-signal intensity to surrounding muscles on T1-weighted (T1W) images, heterogeneously high signal

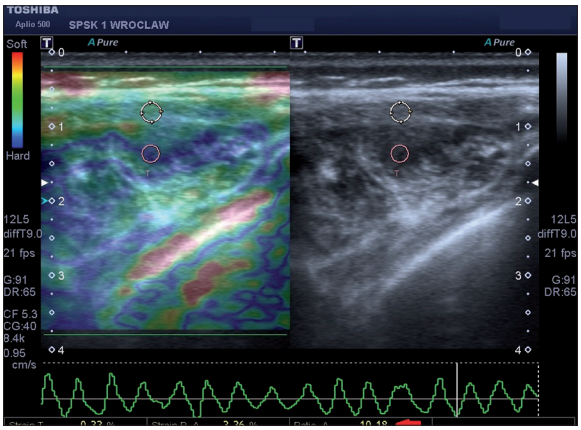

Figure 3. Ultrasound elastography of the right axillar area. A B-mode image reveals a large heterogeneous hypoechoic mass between the surrounding muscles. Elastography reveals increased stiffness in comparison to the surrounding tissue – the mass is about ten times harder than the reference muscle (red arrow).

intensity on T2-weighted (T2W) images, and showed heterogeneous contrast enhancement after intravenous gadolinium administration. On diffusion-weighted imaging (DWI) the mass presented the highest signal intensity. Apparent Diffusion Coefficient (ADC) was also measured. The Region of Interest (ROI) was placed in several areas of the pathological mass. ADC values ranged from 1.63 to 2.11 $\times 10^{-3}$ mm^{-2}/s. There are no unambiguous cut-off values that would allow to differentiate between malignant soft tissue tumors and benign desmoid tumors. However, the study by Oka et al. revealed that ADC values of over 1.3 $\times 10^{-3}$ mm^{-2}/s were present only in lesions belonging to the latter pathology [8]. Hence, ADC values measured in our patient strongly indicated benign character of the mass. MRI examination revealed also a few smaller lesions, similar in morphology on T1W and T2W images, located in the neck and lumbar region on the left side, which were not visible on CT scans. Only on DWI sequences it was evidently noticeable that those small pathological foci were a part of a large infiltrating mass (Figure 2). Surgical treatment was not possible.

To get more information about the infiltration of tissues, the lesions were examined with elastography which was performed with Toshiba Aplio 500. Unfortunately, due to patient's excitability and lack of cooperation it was impossible to precisely define margins of infiltration. However, elastography revealed visibly higher stiffness of the mass in comparison to uninvolved tissues (Figure 3).

The patient was eventually qualified for chemotherapy. The effects of treatment were assessed using MRI.

Discussion

Aggressive fibromatosis has been defined as tumor-like fibroblastic proliferation of unclear etiology presenting a tendency to infiltrate surrounding structures and to recur after surgical resection without cytologic features of malignancy and tendency to metastasize. [3] The main differential diagnosis includes: benign soft tissue lesions such as fibroma, neurofibroma, malignant soft-tissue sarcoma, and extranodal lymphoma [6].

In pediatric patients it is more likely to suspect malignancy due to higher prevalence of malignant diseases and rare cases of fibromatosis in childhood [9].

Typically, aggressive fibromatosis lesions are isointense on T1-weighted images with visible contrast enhancement, and show heterogeneously high signal intensity on T2-weighted images and on fat-saturation sequences. Calcifications are uncommon. [6].

In the presented case lesions have typical appearance in MRI examination. Presence of calcifications and very young age of the patient are the features that discern this case.

However, during the analysis of the MRI examination it was observed that DWI seems to be more sensitive in bordering the infiltration than T2W images and T2W fat-suppressed images (Figure 2). Therefore, we propose consideration of DWI images analysis before potential surgical treatment. ADC values can be useful in differentiating between malignant and benign tumor [8].

In our case the role of elastography in diagnosing an infiltration remains undefined due to the lack of patient's cooperation. However, some scientific reports revealed usefulness of elastography in diagnosing and monitoring infiltrations of the muscles or spastic conditions [10].

Conclusions

In conclusion, eventual diagnosis of aggressive fibromatosis is based on histopathological examination. However, it is an important condition that should be included in a differential diagnosis of soft-tissue masses in diagnostic imaging. Radiologists should be careful especially in defining margins of infiltration in case of potential surgical treatment.

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