

Case Report

Aggressive fibromatosis of the leg and sacrococcygeal region: a report of two cases

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Abstract: Aggressive fibromatosis is a rare soft tissue tumor that composes of myofibroblasts that arise from musculoaponeurotic structures. It usually affects the abdominal wall but may be also found in other less common sites including the head and neck, submucosa of the oral cavity, spinal, haunch and limbs, especially, the limbs and sacrococcygeal region are rare locations. We described two cases of aggressive fibromatosis. One was 3-year-old girl with aggressive fibromatosis arising from the right leg region. The other was 20-year-old female arising from in the sacrococcygeal region. They were resected with satisfied results. Pathological examination showed that they were composed of fibroblasts, fibrocytes and bundles of collagen fiber. The aggressive fibromatosis, although rare, should be differentiated from some other soft tissue tumors with similar histological features and different localizations of intra-abdominal, abdominal wall and extra-abdominal.

Keywords: Aggressive fibromatosis, leg, sacrococcygeal, pathology

Introduction

Aggressive fibromatosis (AF) is also known as desmoid tumor, which was fully described by Stout in 1954 [1]. It is a rare soft tissue tumor and composes of myofibroblasts that arise from musculoaponeurotic structures. The localization of aggressive fibromatosis is mostly in the abdominal wall but may be also found in other less common sites including the head and neck, submucosa of the oral cavity, spinal, haunch and limbs, especially, the limbs and sacrococcygeal region are rare locations of aggressive fibromatosis [2]. Preoperative diagnosis of AF is difficult because of atypical symptom except a rapidly enlarging mass, and it mainly depends on postoperative pathological examination. In view of lacking cell atypia, mitosis and tissue necrosis, AF is deemed to a low grade and non-metastasized tumor. However, it tends to local recur without a complete resection including the edge of the tumor, so long-term follow-up is recommended. We herein report two cases of aggressive fibromatosis

and then discuss the possible differential diagnosis.

Case presentation

Case 1

A 3-year-old girl was admitted to the Department of Orthopedics with a soft tissue mass in the right leg region for 3 months. The lesion had a rapidly progressive course with mild right leg pain over the next 1 month. There was no history of trauma, pain or discharge related to the swelling. He denied any relevant previous or family history. On physical examination, there was a nontender, soft, rounded, mass measuring approximately 6 cm×4 cm×2 cm. No red and swollen were noted. Magnetic resonance imaging (MRI) of the soft tissue mass revealed long oval mass lesion growing along the fibers of the right leg, T1-weighted showing an equisignal, Diffusion weighted imaging (DWI) showing T1-weighted and T2-weighted low signal image of small dot signal (**Figure 1**). Enhancement

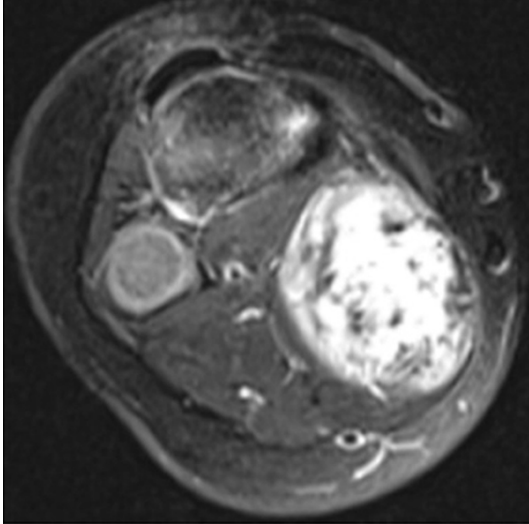


Figure 1. Magnetic resonance imaging (MRI) revealed T1-weighted showing an equisignal, Diffusion weighted imaging (DWI) showing T1-weighted and T2-weighted low signal image of small dot signal.

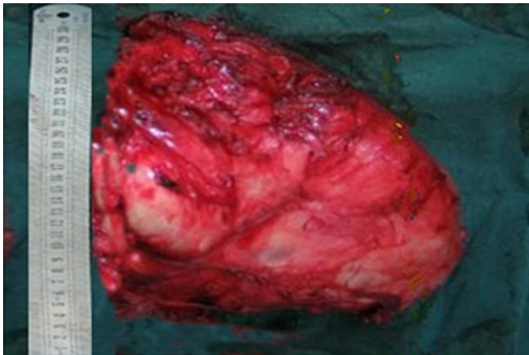


Figure 3. Gross appearance of the resected specimen, which measured 6 cm×4 cm×3 cm in the right leg region.

scan showed lesion was significantly intensify, while the low signal image of small dot signal was not (**Figure 2**).

At laparotomy, the solid mass was located deep fascia and the calf muscles, and gross total resection was performed. The excised mass was approximately 6 cm×4 cm×3 cm (**Figure 3**). Pathological examination, the resected specimen was composed of haphazardly arranged, long, slender spindle cells in a collagenous matrix and sparse fibroblast (**Figure 4**). Based on pathological examination, the diagnosis of aggressive fibromatosis was rendered. Postoperative course was uneventful; the patient had an uncomplicated recovery and

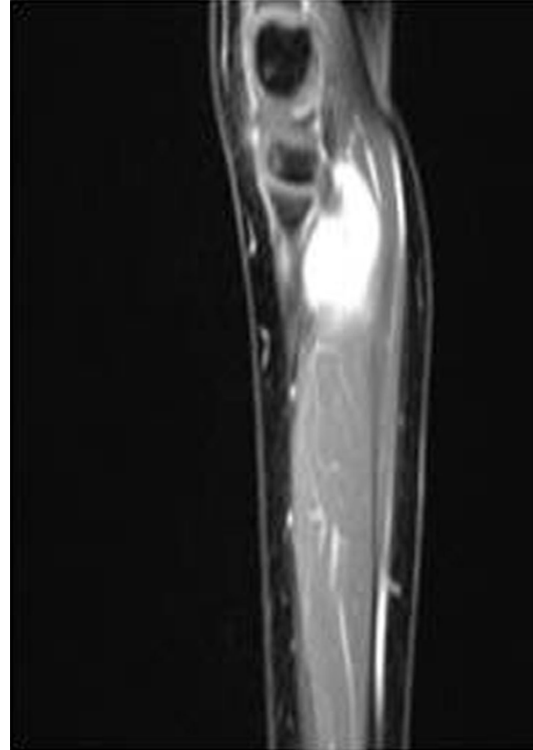


Figure 2. Magnetic resonance imaging (MRI) revealed enhancement scan showed lesion was significantly intensify.

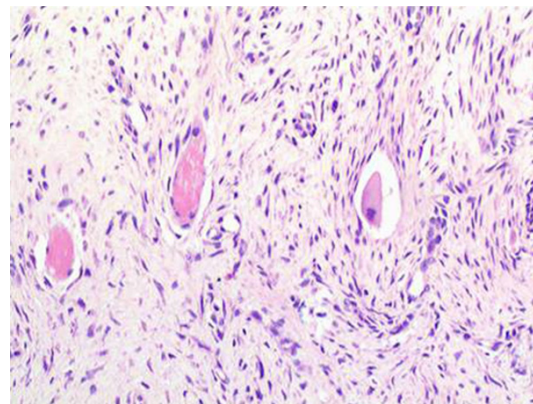


Figure 4. Histologic features of the lesion showed the tumor was composed of haphazardly arranged, long, slender spindle cells in a collagenous matrix and sparse fibroblast (HE×200).

was free of disease at 2 years postoperative follow-up.

Case 2

A 20-year-old female was admitted to the Department of Orthopedics with a soft tissue

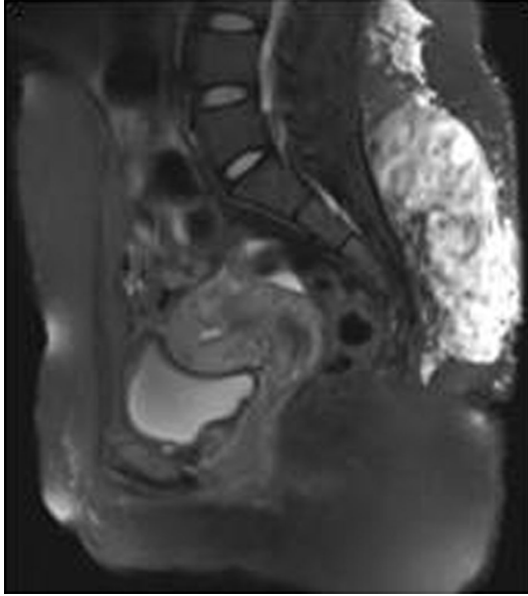


Figure 5. Magnetic resonance imaging (MRI) revealed T1-weighted and T2-weighted showing equi-signal and small dot and strip signal.

mass in the sacrococcygeal region for 8 months. Firstly, the size of the tumor just as bean, the lesion had a rapidly progressive course with waist pain over the next 1 month. There was no history of trauma, pain or discharge related to the swelling. He denied hematochezia, defecate abnormally and rectal irritation. On physical examination, the activity of limbs was well. We could see a promontory on the skin of sacrococcygeal region. there was a slight tenderness, soft, rounded, mass measuring approximately 5 cm×3 cm×1cm. Magnetic resonance imaging (MRI) of the soft tissue mass revealed there was an abnormal crumby signal and the muscles of sacrococcygeal region was consistent with long axis. T1-weighted and T2-weighted showing equisignal and small dot and strip signal (**Figure 5**). Enhancement scan showed lesion was rich in blood and invaded the adjacent blood (**Figure 6**).

At laparotomy, the solid mass was identified in sacrococcygeal region. We tried our best to resect the lesion integrally. The excised mass was approximately 5 cm×2 cm×2 cm (**Figure 7**). Pathological examination, the resected specimen was composed of fibroblast, which haphazardly arranged in bunch or glandular. Composing spindle cells in a collagenous matrix

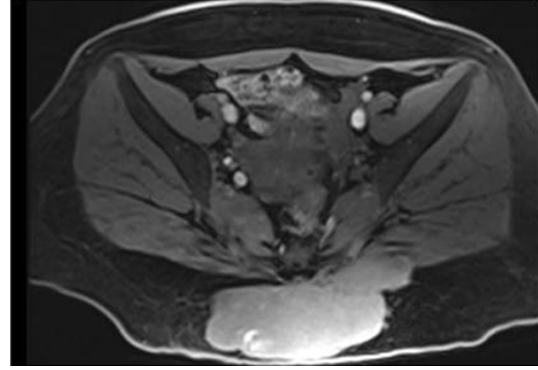


Figure 6. Magnetic resonance imaging (MRI) revealed enhancement scan showed lesion was rich in blood and invaded the adjacent blood.

(**Figure 8**). Based on pathological examination, the diagnosis of aggressive fibromatosis was rendered. Postoperative course was uneventful; the patient had an uncomplicated recovery and was free of disease at 1 year postoperative follow-up.

Discussion

Aggressive fibromatosis (AF) is a rare tumor of deep-seated musculoaponeurotic origin that primary affects the abdominal. It may be also occur in other less common sites including the head and neck, submucosa of the oral cavity, spinal, haunch and limbs. AF was first described by Dupuytren in 1839. However, it was not fully recognized until 1954 by Stout.

In regards to the clinical presentation of normal mitosis and nonmetastasis, AF was considered as benign fibrous tumors in the past, but we found the AF were often poorly defined and had a high potential for recurrence and local invasion, such as muscles and neurovascular structures [3]. The incidence of AF is estimated to be about 2-4 per 1000,000 per year and no obvious morbidity of sex ratio. It can occur at any age, but are more commonly occurring between ages of 6 and 15, and again around the age of 40 [4]. The etiology remains unclear, but it may have an association with genetic predisposition, trauma, familial adenomatous polyposis, endocrine factors and Gardner's syndrome [5]. Clinical presentation is asymptomatic except a slowly enlarging painless soft tissue mass. The patient 1 and 2 all suffered a rapidly progressive lesion with asymptomatic over the next 3 and 8 months, Magnetic resonance imaging



Figure 7. Gross appearance of the resected specimen, which measured 5 cm×2 cm×2 cm in sacrococcygeal region.

(MRI) image suggested fibromatosis. We resected the mass, postoperative pathology diagnosed resected specimen as aggressive fibromatosis.

Iconography examination plays an important role in diagnosing AF. Generally speaking, when MRI images show most of the lesion infiltrate to one or more muscle, the edge of some which are lobulated and clawed, others are quasi-circular and fusiformis. The MRI signal and degree of reinforcement have something with components of the cells, collagenous fiber and the interstitial blood vessels [6]. What is more, the most important feature of MRI is that the area of collagenous fiber in lesion seen on T1WI and T2WI are low signal, however, the area of the cells in lesion seen on T1WI is isointensity signal, on T2WI is high signal, and they usually appeared with obvious enhancement. Besides, CT imaging showed inhomogeneous mild enhancement after contrast inject, while some could enhanced intensely, especial on the delay scan [7].

Although imaging examination will certainly help for AF, but diagnosis of AF mostly depend on postoperative pathological histological fea-

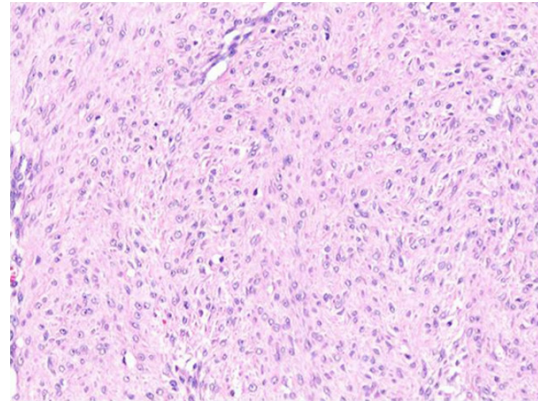


Figure 8. Histologic features of the lesion showed the tumor was composed of fibroblast in bunch or glandular and spindle cells in a collagenous matrix. (HE×200).

tures and immunohistochemical evaluation. Routine pathobiology feature of AF is composed of fibroblasts, fibrocytes and bundles of collagen fiber, which is invasive growth without diolame. what is more, Borders of fibroblasts of AF is clear and no atypia, they list in beam pattern. Immunohistochemical staining show that Vim, Bcl-2, CD99, SMA, CD34 etcetera are positive [8].

AF should be differentiated from by the localization that is intra-abdominal, abdominal wall and extra-abdominal [9]. First of all, AF which occurs in extra-abdominal sites should be differentiated from some other soft tissue sarcoma: fibrosarcoma, liposarcoma, or malignant fibrous histiocyteoma (MFH). Fibrosarcoma is most likely to be confused with AF in some other soft tissue sarcoma. For one thing, we can find the edema area in the edge of Fibrosarcoma, for another, Borders of fibrosarcoma of AF is unclear and inhomogeneous signal. Other soft tissue sarcoma what should be distinguished from AF is Liposarcoma. There is full envelope and edema around the muscles, especially, we can find fat in liposarcoma [10]. MFH is another soft tissue sarcoma that may be confused with AF. The high morbidity of MFH is the aged, but AF is frequently found in the young. Boundary of MFH is not clear and we can find edema area around the MFH, which are inhomogeneous signal in CT imaging [11].

Secondly, AF should be differentiated from some other tumors of abdominal wall. Among the tumors of abdominal wall that can be con-

fused with AF are neurogenic tumor and hematoma after trauma. Neurogenic tumors which have a clear demarcation between the surrounding tissues distribute mainly along the nerve root, and some of them are calcific. Hematoma after trauma are local masses after trauma. The density of traumas are associated with the time of trauma [12].

At last, there is need to make differential diagnosis of AF in relationship to the intra-abdominal, such as gastrointestinal stromal tumors (GISTs), some soft tissue sarcoma, lymphoma. GISTs mainly occur in middle-aged and old people. We can see capsule, necrosis and calcification in the larger GISTs that connected with intestines. CT imaging showed intense enhancement after contrast inject [13]. Making different with soft tissue sarcoma is the same as the AF which occur outside of abdominal. Lymphoma, the multiple nodule fusion or lobulated, usually have no regular shape. It is easy to see enlarged lymph nodes in peritoneum and mesenteric vessel embraced by lymph nodes [14].

Although aggressive fibromatosis is benign histopathologic features, because of infiltrating growth pattern and the presence of adjacent structures such as nerves and vessels, treatment of AF is complete surgical excision. But local recurrence may occur, when the excision comes to adjacent structures. So it is hard to excise completely. Therefore, the follow-up as long as possible is necessary and the adjuvant chemotherapy or radiotherapy are good choice besides surgical excision [15]. The patient reported were all free of disease postoperative follow-up.

In conclusion, there is no clear diagnostic criteria for aggressive fibromatosis, Its diagnosis should be made by their microscopical, histopathological features and iconography examinations. Aggressive fibromatosis should be considered in the differential diagnosis of lesions that occur intra-abdominal, abdominal wall and extra-abdominal such as Fibrosarcoma, Liposarcoma, Neurogenic tumor and lymphoma.

Disclosure of conflict of interest

None.

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