



Large Multicentric Synchronous Extra-Abdominal Fibromatosis of the Leg and Foot: A Case Report

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Abstract

Extra-abdominal fibromatosis is an uncommon, benign locally aggressive fibrous soft-tissue tumor that usually occurs in the shoulders, chest wall, back, thigh, and head and neck affecting the young adult population. It is commonly located in the subcutaneous tissue and may infiltrate the adjacent skeletal muscles. We hereby report a rare case of a large extra-abdominal fibromatosis of the leg and foot in a 38-year-old woman. The patient presented with a large voluminous lesion clinically and on imaging, which was difficult to diagnose. Magnetic resonance imaging (MRI) was very helpful in diagnosing the lesion. It revealed a large relatively well-defined, lobulated hypointense mass in the posterior compartment of the leg with extension into the lower thigh and foot with local infiltration into the gastrocnemius and soleus muscles. An incisional biopsy was performed, and the mass was diagnosed on pathological examination as a spindle-shaped fibroblast proliferation suggesting extra-abdominal fibromatosis.

Keywords

- ▶ desmoids
- ▶ extra-abdominal
- ▶ fibromatosis

Introduction

In 1838, Muller had described the Greek term “desmoid,” which means tendonlike.¹ Fibromatosis or desmoid tumors are rare soft-tissue tumors accounting for 0.03% of all tumors and less than 3% of all soft-tissue tumors. It affects 1 to 2 per 500,000 people worldwide. Incidence of desmoid tumors is approximately 2 to 4 per 1 million population per year in the general population.^{1–3} They commonly occur in the second to third decade of life, and there is female predilection.

Desmoid tumors are usually small and not attached to the skin and subcutaneous tissue. Sometimes, they may be attached to the deeper structures and may be painful. This article will summarize the features of desmoids that radiol-

ogists should be aware of, specifically common locations, complications, and treatment-related changes. Here, we present a rare case of extra-abdominal fibromatosis of the leg and foot, and it appears to be the largest lesion in the literature to the best of our knowledge.

Case History

A 38-year-old woman presented with an 11-year history of a very slow-growing hard lobulated nodular growth on the posterior aspect of the leg and the dorsum of the foot. The growth extended superiorly up to the lower thigh region. The physical examination showed a firmly fixed mass with areas

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Fig. 1 Clinical photograph of a 38-year-old woman showing nodular swelling on the posterior aspect of the leg and foot. Skin is thin and shiny with areas of skin discoloration.

of ulceration in the foot region. The skin had become thin and shiny in certain parts (► **Fig. 1**).

There was no history of any trauma. She did not have any medical history or similar illness in family members. The other laboratory investigations were within normal limits. Radiography revealed a relatively well-defined, lobulated, increased soft-tissue density lesion seen in the posterior aspect of the leg and foot with extension into the posterior aspect of the lower thigh. No obvious bony erosion was seen (► **Fig. 2**). Ultrasonography revealed an inhomogeneously hypoechoic lobulated soft-tissue lesion in the muscular plane and showed posterior acoustic shadowing at places (► **Fig. 3**). Color Doppler study revealed areas of increased vascularity in the leg and foot. On computed

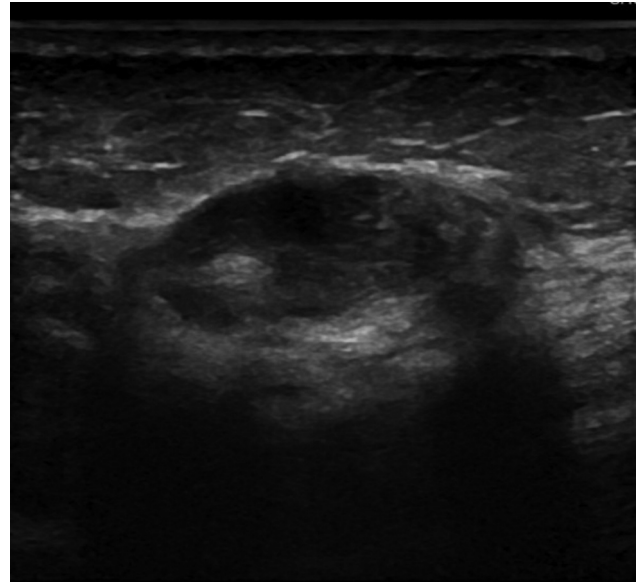


Fig. 3 Ultrasound (GE Logiq S8, linear transducer, frequency 12 MHz) of the right leg demonstrates an inhomogeneously hypoechoic lobulated soft-tissue lesion in the muscular plane with posterior acoustic shadowing at places.

tomography (CT) evaluation, the lesion was heterogeneous with variable attenuation seen in the posterior compartment of the leg involving the gastrocnemius and soleus muscles. There was another heterogeneous lesion with ulceration noted in the foot region causing erosion of cuboid, fourth, and fifth metacarpals (► **Fig. 4**). On magnetic resonance imaging (MRI), the lesion measured approximately 9.7 × 5.2 × 45.0 cm. It was heterogeneous and predominantly hypointense on T1 and T2-weighted (T2W) images as well as STIR sequences with small isointense signal intensities seen within (► **Fig. 5**). The lesion showed postcontrast enhancement in the isointense areas. It was seen



Fig. 2 (A) Anteroposterior (AP) and (B) lateral radiographs show a well-defined, lobulated, increased soft-tissue density lesion in the posterior aspect of the right leg and foot with extension into the posterior aspect of the lower thigh. No obvious bony erosion or calcifications.

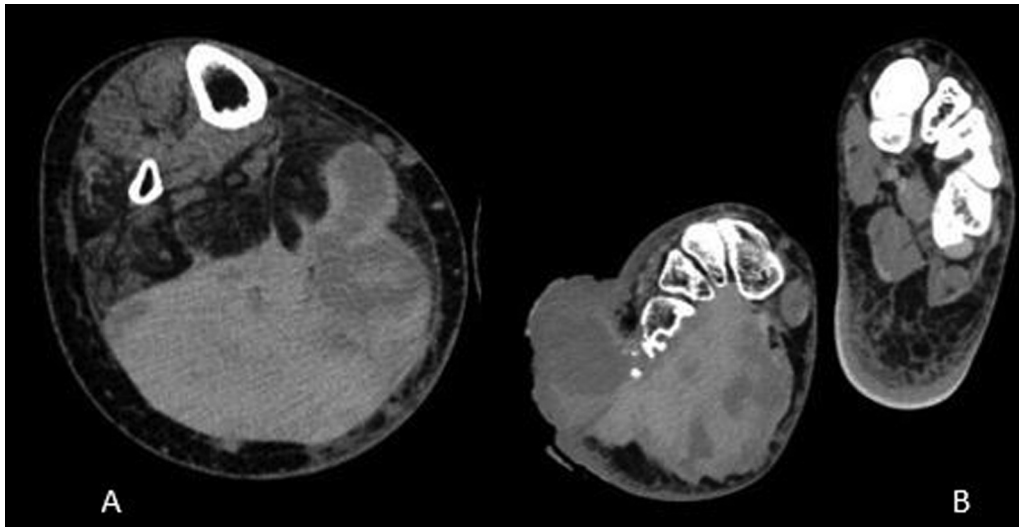


Fig. 4 (A) Nonenhanced computed tomography (CT) section (Phillips ICT 256 slice CT scanner) through the upper right leg demonstrates a heterogeneous hyperdense lesion in the posterior compartment of the leg involving the gastrocnemius and soleus muscles with few hypodense areas. (B) Axial section through the foot shows another heterogeneous lesion with ulceration in the foot region and erosions involving the cuboid, fourth, and fifth metacarpals.

predominantly in the posterior compartment of the leg and also extending into the distal thigh in subcutaneous and intermuscular planes with the involvement of the gastrocnemius and soleus muscles. The rest of the posterior compartment muscles showed atrophy and increased signal due to fatty infiltration. The tibia, fibula, and distal femur showed normal signal intensity without any erosions or extension into the knee joint. No collection in the knee joint was noted. There was another similar intensity lesion with an approximate size of $6.9 \times 6.4 \times 10.4$ cm seen involving the plantar aspect of the foot extending up to the ankle joint. The lesion was extending exophytically into the lateral aspect with loss of skin and soft tissue. The lesion was causing erosions of the cuboid and the base of the fourth and fifth metatarsals. There was no continuity between the two lesions. Then, the patient underwent a USG-guided biopsy from the foot and leg region, and a histopathological examination was performed. The lesion showed benign proliferation of spindle cells with areas of myxoid tissue and no malignant cells were noted (**Fig. 6**). The patient then underwent a surgical resection and was referred for local irradiation.

Discussion

Extra-abdominal desmoid fibromatosis, also called deep or aggressive fibromatosis, musculoaponeurotic fibromatosis, the extra-abdominal desmoid tumor, is an uncommon mesenchymal neoplasm. These tumors can be intra-abdominal, in the abdominal wall, or extra-abdominal, and extra-abdominal desmoid fibromatosis usually affects the shoulder, chest wall and back, thigh, and head and neck.⁴ The disease is rare in the leg and foot. These are locally invasive without the capacity for distant metastases. Local recurrence and adjacent organ involvement are important causes of morbidity and mortality. The incidence of desmoid tumors in the general population is 2 to 4 cases per 1 million per year, with a slight female preponderance and peak incidence in the third and fourth decades of

life.⁵ These tumors behave more aggressively in younger patients, with very high recurrence rates up to 87%.⁶ They present as a firm mass, particularly when extra-abdominal. They have spiculated infiltrative margins and are typically adherent to the adjacent structures.

Desmoid tumors or fibromatoses are broadly classified as superficial (involving the superficial fascia) and deep (involving the deep musculoaponeurotic system). Deep fibromatosis is of three types: extra-abdominal, affecting the extremities (shoulder, chest, calf, and thigh); abdominal, affecting the abdominal wall; and intra-abdominal, affecting the small bowel mesentery. The intra-abdominal type has more syndromic associations like familial adenomatous polyposis and Gardner's syndrome.⁷⁻⁹ Tumor can be multicentric in 10 to 14% of cases. Multicentric extra-abdominal desmoids are rare and can be metachronous or synchronous.^{10,11} They are locally aggressive, and recurrence is common, but they do not metastasize.⁸ Recurrence is commonly seen in young patients presenting with less than 30 years of age. The exact etiology is not known, but multiple factors such as trauma, postsurgical, genetic or hormonal imbalance are known to be its causes of occurrence. There are various imaging modalities available for diagnosis of desmoid tumor. Radiographs and CT scans are of limited use. They demonstrate the soft-tissue shadow if the lesion is large and calcifications on radiograph and CT. MRI is the modality of choice due to excellent soft-tissue differentiation and tells us about the accurate extension and invasion of the lesion into the surrounding planes. On MRI, the lesion usually appears as a lobulated heterogeneous mass involving the intermuscular plane and fascia with areas of bandlike low signal intensity on T1 and T2W images. The closest differential diagnosis could be malignant soft-tissue sarcoma or alveolar soft part sarcoma (ASPS). But in general, sarcomas are very vascular lesions and demonstrate the intratumoral and peripheral flow voids, central area of necrosis, appear

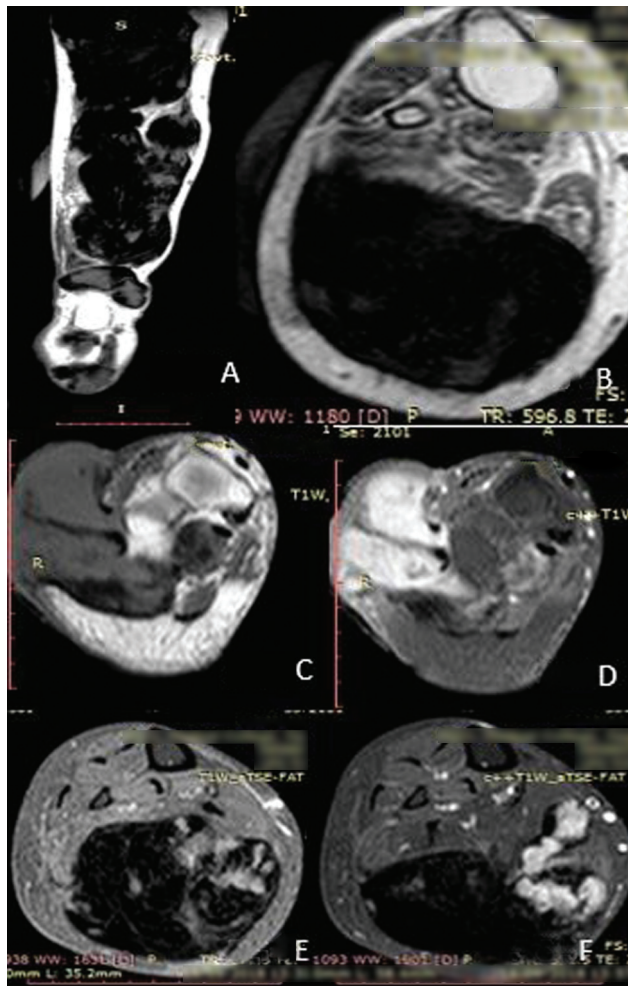


Fig. 5 Magnetic resonance imaging (MRI; Philips Achieva 1.5 T) shows a large heterogeneous lesion predominantly hypointense on (A) T1-weighted (T1W) and (B) T2-weighted (T2W) in the posterior compartment of the leg extending into the distal thigh in the subcutaneous and intermuscular planes with the involvement of the gastrocnemius and soleus muscles. (C) Precontrast T1W and (D) postcontrast T1W images show another similar enhancing lesion involving the plantar aspect of the foot extending up to the ankle joint with the involvement of the skin and soft tissue. (E) short tau inversion recovery (STIR) sequences with small isointense signal intensities seen in the leg lesion shows postcontrast enhancement (F) in the isointense areas.

predominantly hyperintense on T1W images, and maintain the facial planes early in the disease.^{12,13} In our case, the lesion did not show any peritumoral or intratumoral flow voids; it involved the adjacent facial planes and predominantly showed hypointensity on all pulse sequences, which is characteristic of fibromatosis.¹²

To the best of our knowledge, lymph node involvement in extra-abdominal desmoids is very rare and not reported in the literature, whereas regional lymph node involvement is more common than distant metastasis in soft-tissue sarcoma. Regional lymph node metastases are seen in 1.75 to 5.9% in soft-tissue sarcoma. Lymphatic spread is more commonly involved in some specific histologic types, such as rhabdo-

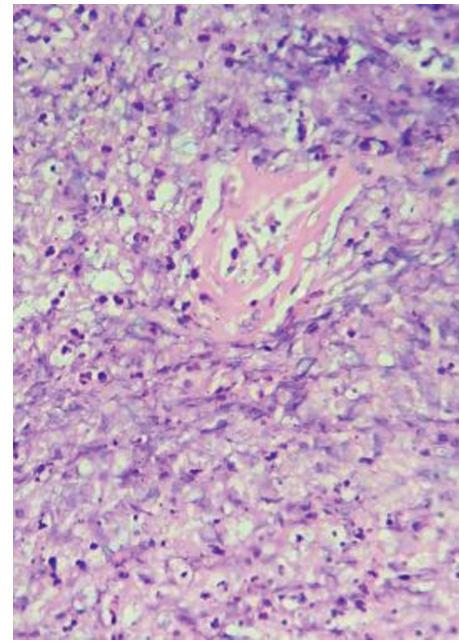


Fig. 6 Ultrasonography (USG) guided biopsy slide photograph showing benign proliferation of spindle cells with areas of myxoid tissue.

myosarcoma, epithelioid sarcoma, clear cell sarcoma, and vascular sarcoma.¹⁴

PET/CT imaging can help us differentiate between metabolically very active tumors like sarcomas and less active lesions like desmoids. Desmoid tumors show a heterogeneous pattern of uptake with standardized uptake values (SUVmax) ≤ 4.8 , whereas sarcomas show very avid uptake with SUVmax greater than 5. ASPS shows SUVmax of up to 9.^{15,16}

Multimodal treatment therapy is suggested for extra-abdominal fibromatosis that involves surgery, hormonal therapy, nonsteroid anti-inflammatory drugs (NSAIDs), and radiotherapy, and they are given alone or in combination. In large lesion amputation and recurrences, excision is also advised. Sometimes, when the tumor is not easily resectable, hormonal therapy with tamoxifen is also given for regression of the lesion.⁸ In summary, we present a rare case of large multicentric extra-abdominal desmoid tumor involving the leg and foot in a female patient. To our knowledge, this is the largest described multicentric synchronous extra-abdominal fibromatosis in the literature. Early imaging of the lesion can prevent morbidity and recurrence in such patients.

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None.

Conflict of Interest
None declared.

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