DESMOID TUMOR : A CASE REPORT

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Extra-abdominal fibromatosis (desmoid tumor) is an uncommon soft tissue tumor. About two-third of cases occur in the extremities. We present a female 43-year-old case of multicentric desmoid tumor.

CASE REPORT

A 43-year-old woman came to the Chulalongkorn Hospital in June 1998 presenting with painless, non tender and slow growing masses at left calf and foot for a year. She had previous surgery of left thigh mass since she was 12 year-old. Physical examination revealed soft tissue mass, 15x10 cm in size at dorsum of left equinous foot, which was fixed, firm consistency and ulcerated on top. The lesion along left calf was composed of multiple small masses, about 1.5 cm. in size and a big mass, 8 cm. in size with the surgical scar on the skin. Neither neurological deficit nor vascular compromise was observed. There was no palpable node at both inguinal areas. The laboratory data were within normal limits. The plain radiographs of the left leg and foot revealed large soft tissue masses at dorsum of left foot and along posterior aspect of left leg. Multiple cortical bony projections of fibula and tarsal bones were identified being spicule-like appearance (Fig. 1 A,B). The tarsal bones showed well defined cystic changes due to pressure erosion (Fig. 2). Three times of tissue biopsies were taken. The first and second specimens revealed chronic inflammation and fibrosis. The last one showed fibroma of tendon sheath. Wide excision was performed subsequently. Pathologically, the lesion was poorly circumscribed and consisted of elongated spindle-shaped cells of uniform appearance which separated by abundant collagen. There was no cellular atypia. The constituent nuclei were small, pale staining, and had one minute nucleoli (Fig. 3 A, B).

DISCUSSION

Fibromatosis (desmoid tumor) refers to locally invasive tumor of connective tissue and its overlying fascia or aponeurosis. The biologic behavior is intermediate between that of benign fibrous lesion and that of fibrosarcoma, although never metastasize.[1-3] According to Enzinger and Weiss,[4] the fibromatoses are classified on the basis of their anatomic location as either superficial or deep. The superficial group includes palmar fibromatosis, plantar fibromatosis, penile fibromatosis and knuckle pads fibromatosis. The deep or musculoaponeurotic fibromatoses include extra-abdominal fibromatosis (aggressive fibromatosis), abdominal fibromatosis, and intra-abdominal fibromatosis.

Extra-abdominal fibromatosis typically presents in young adult between puberty and 40 years of age, with a peak incidence between 25 and 35 years. Reports in the literatures indicate

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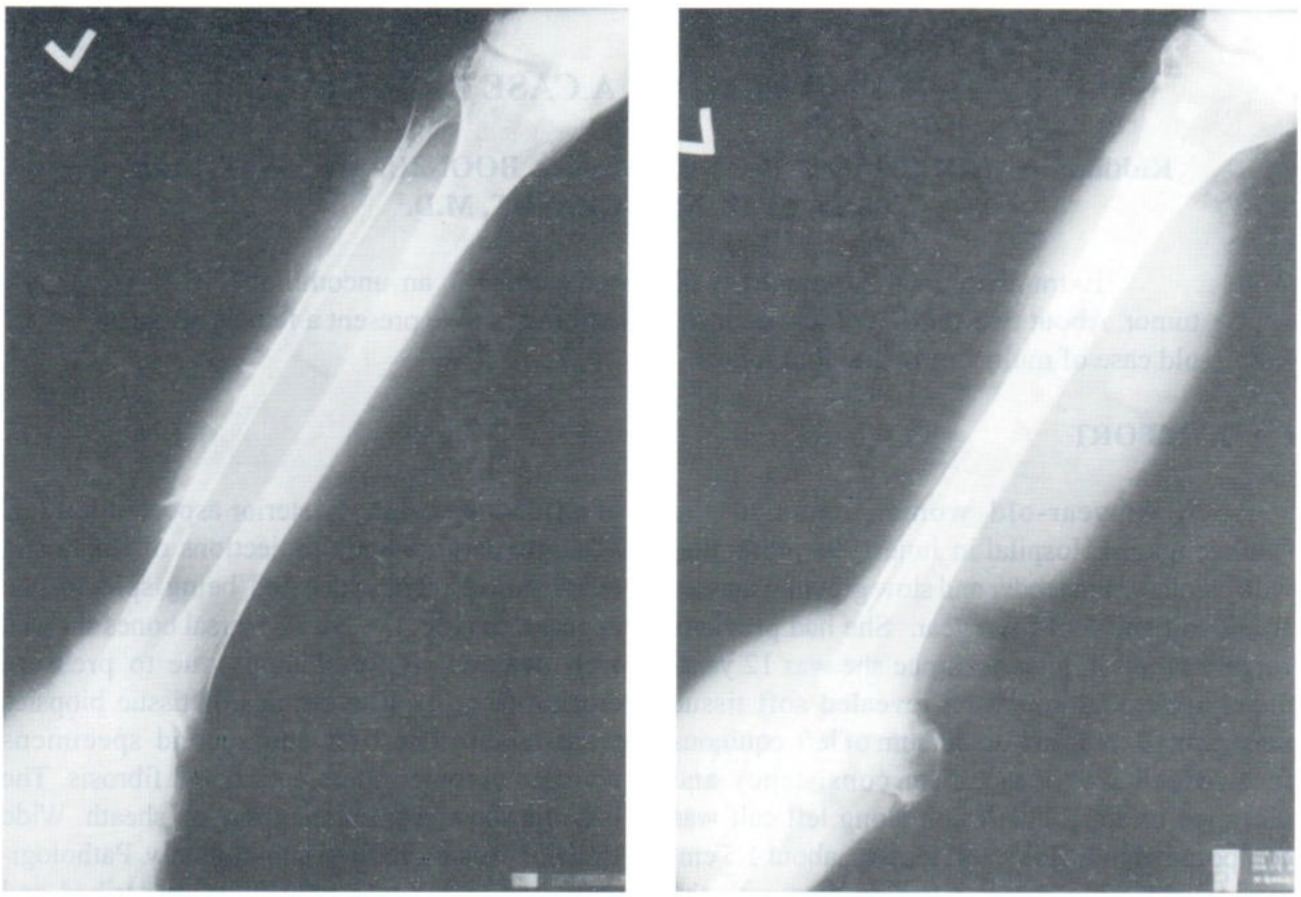

1A 1B

Fig. 1 A,B Plain radiographs of left leg (AP, lateral) showing large soft tissue masses at dorsal foot and calf with cortical spiculation of tibula and tarsal bone

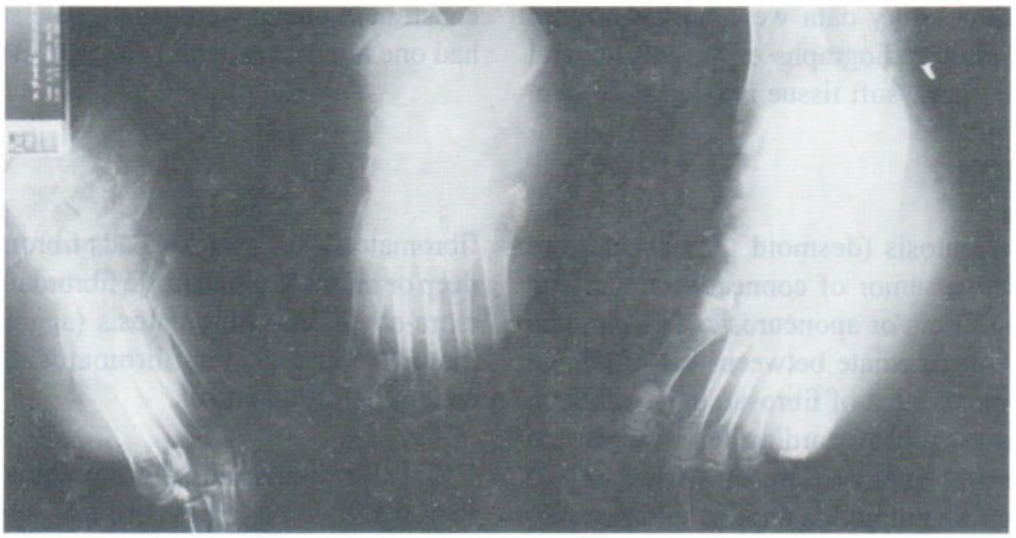

Fig. 2 Plain radiograph left foot (AP, lateral, oblique) showing soft tissue mass of dorsal foot with well defined cystic changes of tarsal bone or pressure erosion.

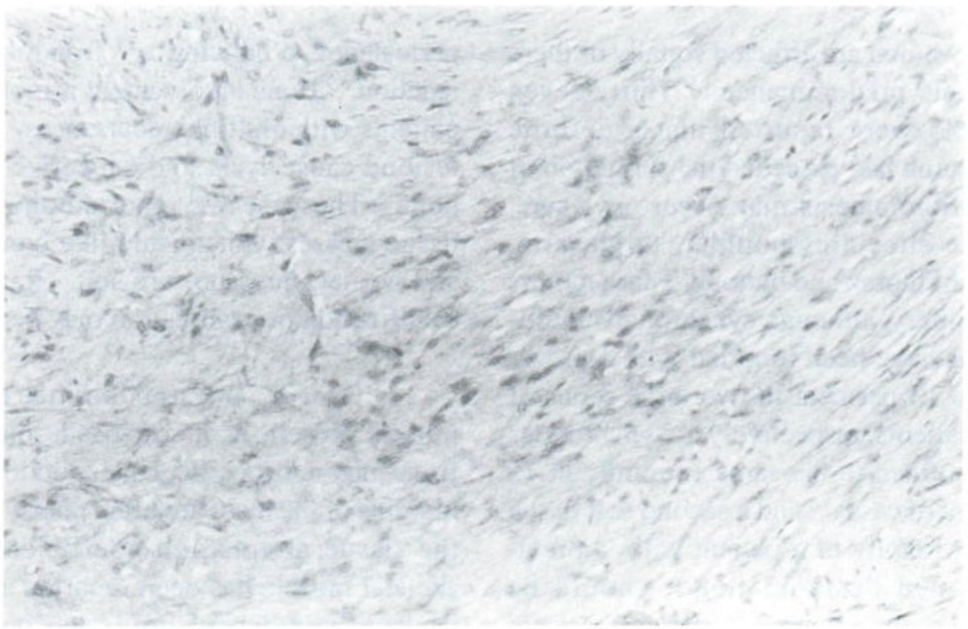

Fig. 3 A Low power picture showing interlacing bundles of fibroblasts separated by large amounts of collagen. (x 200)

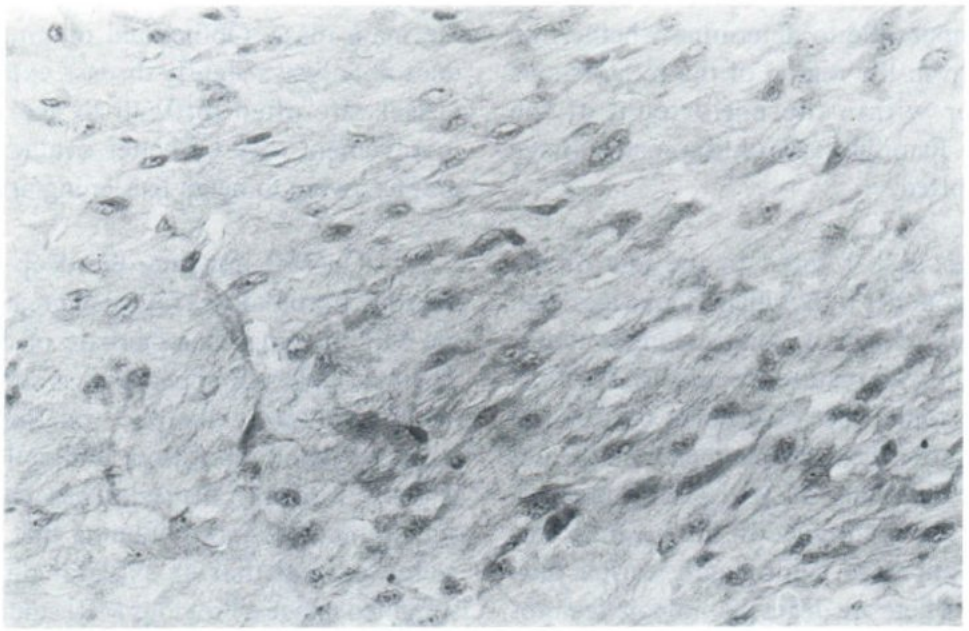

Fig. 3 B High power picture showing vesicular nuclei with minute nucleoli, rather indistinct cytoplasm, and interstitial collagen. (x 400)

that men and women are affected equally or there is 1.8 :1 female predominance.[1,5] Thirty seven patients (19%) were reported that a definite episode of trauma had preceded the formation of a mass. The fibromatosis may occur anywhere. The principle sites are shoulder, thigh, arm, posterior part of thorax and buttock. These tumors are usually solitary, although synchronous multicentric lesions have been reported with a prevalence of 10 to 15 per cent in two large series of 192 and 110 patients, respectively.[5-6] Synchronous lesions are confined to the same extremity in 75 to 100 per cent of cases[5-6] and a second soft tissue mass in the extremity of a patient with a previously confirmed desmoid tumor should be regarded as a second desmoid tumor until proved otherwise. Very rarely the lesion may be juxtacortical. Dong et al[7] reported a juxtacortical lesion in the forearm of a 14-year-old boy, noting that it was impossible to determine whether the tumor arose from the region of the interosseous membrane or within the periosteum of the adjacent bone. Familial cases of fibromatosis have also been reported.

Prognosis is related to the age of the patient, with younger individuals (those less than 20 to 30 years of age) having a longer tumor activity and a higher recurrence rate.[5] 68% of the patients were found that recurrence occurred at about 2 years after the first treatment and greater risk in female older than 30 year-old.[5] Romero et al[8] noted that the recurrence in the juvenile was multiple and appeared significantly earlier than adult patients. There are different biologic features between juvenile and adult patients with histologically same desmoid tumor.

Radiographs are usually nonspecific but roughly estimate of the size and location of the mass. Approximately 6 to 37 per cent of patients have evidence of bone involvement which is usually a pressure erosion, a scalloping without invasion or destruction or stimulation of the periosteum, producing a " frondlike" periosteal reaction.[5,9] Bone involvement is more common in patients with multiple recurrences.[5] Juxtacortical lesions cause lysis and saucerize the adjacent bone.[7] The case we reported showed the soft tissue masses with spicule-like bony projections and cortical pressure erosion. Rare cases with calcification or ossification may be seen.

On CT, extra-abdominal fibromatoses images present as a soft tissue mass, which is frequently nonspecific. Unless outlined by fat, the margins of the mass are often poorly defined,[3] and the tissue attenuation coefficient relative to skeletal muscle has been reported as hypodense, isodense and hyperdense.[2-3] Lesion is enhanced with intravenous contrast with better delineation.[2-3] CT afford the opportunity to discern the proximity of bone and neurovascular structure to the main mass. Obliterated intermuscular plane does not always signify disease extension. Small vessels are often difficult to see. Subtle bone involvement may be better evaluated on radiographs owing to beam hardening artifact.[3]

On MR imaging, the deep musculoaponeurotic fibromatoses revealed great variability of the MR imaging characteristic of the lesions.[3,11] Their variable MR appearance is similar to that of other soft tissue lesion, and this variability reflected the composition and cellularity of the lesion.[11] Typically, the lesion has a heterogeneous signal intensity approximating that of fat on T2-weighted and that of skeletal muscle on T1-weighted images. Previous MRI studies had shown that the signal characteristics of desmoids most closely resemble those of malignant neoplasm such as heterogeneity, poor margination and neurovascular encasement. Mass had areas of low signal intensity consistent with fibrosis on both T1- and T2 weighted images and a signal intensity in the non fibrous portion of the tumor that is less than of fat on T2 weighted image, desmoid should be a primary diagnostic consid-

eration.[12] Fibromatoses typically demonstrate moderate to marked enhancement following administration of the intravenous Gd-DTPA, with enhancement corresponding to the cellular portions of the lesion.[13] MRI provides clear delineation of abnormal tissue and its relation to adjacent vascular and nervous structures suggested that it is more useful than other currently available imaging methods in preoperative staging, planning the surgical approach and determining the potential for limb salvage. However, MRI can be difficult to distinguish reactive edema from malignant lesion on the basis of relaxation time determinations or other characteristics unique to MRI. Overall failure of MRI and CT in diagnosis is that of calcification, ossification and periosteal reaction are less easily evaluated than on plain radiographs.[10]

In conclusion we present an uncommon case of multicentric desmoid tumor with possibly recurrence as correlated with of previous surgery of thigh mass. Only plain radiograph was performed showing large soft tissue mass with pressure erosion and marked bony spiculation. Since no neurovascular symptom was evident so further CT or MRI was unnecessary for additional information owing to limited use of such studies as described.

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