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Magnetic resonance appearance of fibromatosis*

A report of 14 cases and review of the literature

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Abstract. We reviewed retrospectively the magnetic resonance (MR) images of 14 soft-tissue lesions of fibromatosis (desmoid tumors) encountered in 11 patients. The lesions were typically inhomogeneous in texture and round to oval in configuration. Margins were well-defined in 78% of the lesions at presentation and were infiltrating in all recurrences. On T_1-weighted spin echo MR images, the predominant signal intensity was either isointense or minimally hyperintense when compared with skeletal muscle. On T_2-weighted MR images the predominant signal intensity was typically intermediate between skeletal muscle and subcutaneous fat or isointense to fat. Linear and curvilinear areas of decreased signal intensity were distributed throughout the lesions on both pulse sequences in 86% of cases. This pattern strongly suggested fibromatosis. Speculation concerning possible etiologies of this appearance are discussed, and the relevant literature on previously reported cases is reviewed.

Key words: Fibromatosis – Desmoid tumor – Soft-tissue tumors – Skeletal neoplasms – Magnetic resonance imaging

Fibromatosis refers to a family of soft-tissue lesions characterized by proliferation of benign fibrous tissue [4]. These lesions demonstrate biologic behavior intermediate between benign fibrous lesions (such as fibroma or fasciitis) and fibrosarcoma [4, 5, 7]. Although histologically benign, they typically show an infiltrative growth pattern on both gross and microscopic inspection, and local recurrence is common.

Magnetic resonance (MR) imaging is rapidly becoming the favored imaging study for evaluating soft-tissue masses. Our review of the MR appearance of 14 lesions of pathologically proven fibromatosis in 11 patients constitutes the basis of this report.

Materials and methods

We retrospectively reviewed the MR images of 11 patients with 14 proven lesions of fibromatosis (desmoid tumors) of soft tissue. This group was selected from 139 patients with soft-tissue masses of diverse etiology evaluated by MRI at our institution.

The diagnosis of fibromatosis was established in accordance with commonly accepted histologic criteria following biopsy in 13 cases. The final patient had fibromatosis previously established by biopsy. This patient presented with extensive recurrence of a chest wall lesion which had now extended into the mediastinum. The clinicians chose treatment by radiation and did not obtain additional tissue.

MRI examinations were performed on either a GE 1.5-T Signa (General Electric, Milwaukee, WI), or a 1.5-T Teslacon (Technicare, Solon, OH) scanner. Scanning sequences included spin echo T_1 (450–700/20–40) and T_2 (1800–2500/60–100) weighted pulse sequences. All lesions were imaged in at least two orthogonal planes and were evaluated for the following features: margin definition, intensity and homogeneity of the signal, and presence or absence of surrounding edema. Additional data recorded included age and sex of the patient at presentation and the specific sites of skeletal involvement.

Results

The 14 lesions of fibromatosis occurred in 11 patients whose mean age was 22 years (range 11–31 years). There were 9 men and 2 women. Three lesions occurred in the upper chest around the shoulder, two occurred in the plantar soft tissues of the foot, and one each occurred in the upper arm, popliteal fossa, pelvis, and medial thigh. There were five recurrent lesions: two in the plantar aspect of the foot and one each in the thigh,

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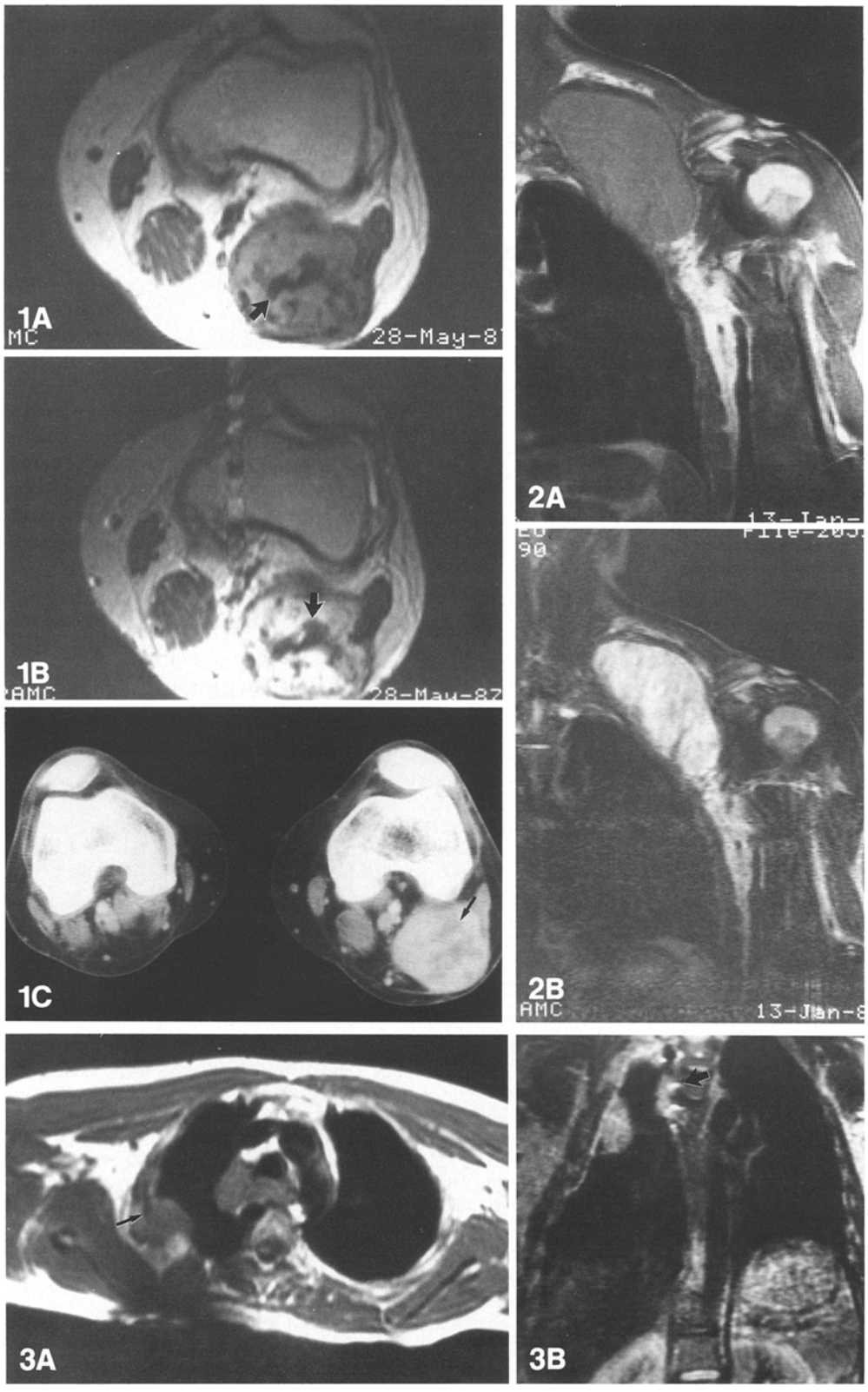

Fig. 1A–C. Fibromatosis of the popliteal fossa, 22-year-old man. Axial T_1-weighted (700/34) (**A**) and T_2-weighted (2000/80) (**B**) MR images show a relatively well-defined, soft-tissue mass in the popliteal fossa. Within the mass, note areas of decreased/absent signal (*arrows*) on both pulse sequences. **C** Corresponding contrast-enhanced axial CT. Note lack of enhancement in the center of the mass (*arrow*) in a distribution corresponding to the signal voids seen on MR

Fig. 2A, B. Fibromatosis of the chest wall, 24-year-old man. Coronal T_1-weighted (700/33) (**A**) and T_2-weighted (2000/100) (**B**) images show a well-defined, soft-tissue chest wall mass. Signal intensity on T_1-weighted (**A**) MR image slightly exceeds surrounding skeletal muscle. Mass is mildly inhomogeneous on T_2-weighted (**B**) MR image with signal intensity greater than fat

Fig. 3A, B. Recurrent fibromatosis of the chest wall, 31-year-old man. **A** Axial T_1-weighted (600/20) image shows a soft-tissue mass infiltrating the chest wall and mediastinum. The signal intensity of the mass approximates to skeletal muscle. Note areas of decreased signal (*arrow*) within the mass. **B** Coronal T_2-weighted (2000/80) image shows the mass (*arrow*) extending over the apex of the lung and into the mediastinum

chest wall, and upper arm. In three patients with recurrence (foot, thigh, and upper arm) the original MR images and histologic slides were available for review. Ten tumors were on the left and four on the right side of the body.

Ten patients (13 lesions) presented with a painless, palpable, soft-tissue mass. The remaining lesion was identified during routine follow-up after resection of a chest wall mass.

The MR images showed margins ranging from well-

defined (Figs. 1A, 1B, 2) to infiltrating (Fig. 3), the latter encountered in all five recurrences and in two of nine primary lesions. The remaining seven lesions had relatively well-defined margins. None of the lesions had surrounding edema.

On T_1-weighted MR images, the predominant signal intensity of all lesions was either isointense with (50%) or slightly hyperintense (50%) to skeletal muscle. Twelve of 14 lesions (86%) demonstrated mild to moderate inhomogeneity including linear and curvilinear areas of signal void interspersed throughout the tumor (7 lesions), peripherally (3 lesions), or centrally (2 lesions) (Fig. 1A). The remaining 2 lesions were relatively homogeneous (Fig. 2A).

On T_2-weighted MR images a similar but more conspicuous pattern of inhomogeneity was seen. In 12 of 14 lesions there was moderate inhomogeneity with areas of signal void corresponding to those identified on the T_1-weighted images (Fig. 1A and B). The remaining two lesions were only mildly inhomogeneous (Fig. 2B). The predominant signal intensities of these lesions were approximately equal to subcutaneous fat (43%), between skeletal muscle and subcutaneous fat (36%), or slightly hyperintense to subcutaneous fat (21%).

Review of histologic sections and MR images showed good correlation between signal intensity and tumor cellularity and collagen content. The tumor with the highest signal intensity on T_2-weighted images was very cellular on histologic examination and had relatively less collagen.

Discussion

Fibromatosis includes a family of soft-tissue lesions characterized by proliferation of benign fibrous tissue composed of uniform, elongated, fusiform or spindle-shaped cells surrounded and separated by abundant collagen [4]. Their biologic behavior is intermediate between benign fibrous lesions and fibrosarcoma although they never metastasize [4, 5, 7]. Synonomous terms include nonmetastasizing fibrosarcoma or aggressive fibromatosis, but these may be misleading since the clinical course of any individual tumor is unpredictable [4].

The most widely accepted classification of the fibromatoses, as noted by Enzinger and Weiss, is presented in Table 1 [4]. The two major categories, superficial and deep fibromatoses, are in turn subdivided according to anatomic location. Superficial fibromatoses are typically slow-growing and small and usually arise from fascia or aponeuroses. Conversely, deep fibromatoses typically grow rapidly and are larger and more aggressive in their biologic behavior. The latter are sometimes termed desmoid tumors, a term initially employed to emphasize their "bandlike or tendonlike" consistency [4].

All fibromatoses show infiltrative growth and are indistinguishable on both gross and microscopic inspection [4]. They are differentiated by anatomic location and clinical presentation rather than morphologic differences.

Growth pattern, relationship with surrounding tissues, and cytologic features are the key histologic criteria that establish the diagnosis of fibromatosis. The growth pattern consists of relatively uniform fibroblasts with bland nuclei proliferating in parallel arrays. There is a variable amount of intermingled collagen, in some cases relatively scant, in others more abundant. In addition, other changes may be seen including myxoid change, focal hemorrhage, vascularity, and focal inflammation, perhaps induced by chronic trauma or injury. The relationship to surrounding tissue (e.g., subcutaneous fat and skeletal muscle) is marked by interdigitating, infiltrative growth. This last-mentioned feature probably causes difficulty in establishing margins of resection, thereby creating a predisposition for local regrowth if excision is incomplete.

Table 1. Classification of the fibromatoses

I. Superficial (fascial) fibromatosis
A. Palmar fibromatosis (Dupuytren's contracture)
B. Plantar fibromatosis (Ledderhose's disease)
C. Penile fibromatosis (Peyronie's disease)
D. Knuckle pads
II. Deep (musculoaponeurotic) fibromatosis
A. Extra-abdominal fibromatosis (extra-abdominal desmoid)
B. Abdominal fibromatosis (abdominal desmoid)
C. Intra-abdominal fibromatosis (intra-abdominal desmoid)
1. Pelvic fibromatosis
2. Mesenteric fibromatosis
3. Gardner's syndrome

Fibromatoses typically present in young adults between puberty and 40 years of age, with a peak incidence between 25 and 35 [4]. They have, however, been reported in children. Reports in the literature indicate that men and women are almost equally affected [4]. The male predominance in our series must be viewed with caution due to the small number of patients and the idiosyncratic referral pattern.

These tumors are usually solitary. Since synchronous multicentric lesions have been reported, a second soft-tissue mass in the extremity of a patient with a previously confirmed desmoid tumor should be regarded as a second desmoid until proven otherwise [12]. Familial cases have also been reported, although none was encountered in this series [4, 12]. Local recurrence is common, occurring in as many as 50% of patients [6]. Recurrence was noted in 5 of our 14 patients (36%) although long-term follow-up is lacking. Fatalities secondary to direct invasion of the chest wall or neck have been reported [2, 6].

As noted in Table 2, scattered examples of fibromatosis (desmoid tumors) have appeared in the MR literature. Sundaram described three cases of aggressive fibromatosis, two demonstrating a decreased signal on T_2-weighted pulse sequences and one showing a paradoxical increased signal [10]. Two of these three tumors were hypocellular and had abundant collagen. The lesion that showed high signal on T_2-weighted MR images had marked cellularity as well as abundant collagen. Sundaram concluded that the combination of marked hypocel-

Table 2. Reported cases of fibromatosis in which the magnetic resonance spin echo imaging characteristics are described

Reference	Lesions (*n*)	Tesla	Description
Aisen et al. [1]	2	0.35	Very low SI on both short and long TR and TE
Chang et al. [3]	2	0.5	No significant differences between T_1 and T_2 values for low grade desmoid, and high grade tumors
Hudson et al. [8]	2	0.15	One well-delineated appearing "black" on spin echo images, one very heterogeneous with bright and dark areas
Petasnick et al. [9]	2	0.5	One equal to SM on T_1 and between SM and fat on T_2, one equal to SM on T_1 and equal to fat on T_2 with lower SI peripherally; both inhomogeneous, with indistinct margins
Sundaram et al. [10]	3	0.15	Paradoxical SI with long and short T_2
Sundaram et al. [12]	4	?	Low SI on long TR/TE sequences
Totty et al. [13]	1	0.35 or 0.5	SI equal to SM on T_1, no T_2 images
Wetzel et al. [14]	1	1.0	Very low signal on both T_1 and T_2

SI, signal intensity; SM, skeletal muscle

lularity and abundant collagen produces decreased signal on T_2-weighted pulse sequences and that the decreased cellularity is of prime importance [10]. Our experience supports Sundaram's finding that the relative cellularity of the tumor is of great importance in determining the signal intensity on spin echo MR images [10].

Aisen described two patients in whom MRI was useful in detecting fibromatosis by demonstrating decreased signal intensity within the lesion on both long and short TR and TE spin echo images [1]. Wetzel et al. reported similar observation in one patient [14]. Hudson et al. also described two cases, one appearing "black" on spin echo images, while the other was heterogeneous and resembled those in our series [7]. Petasnick et al. reported two inhomogeneous soft-tissue masses with indistinct margins and infiltration of adjacent muscle and fat. The signal intensity of one lesion approximated skeletal muscle on T_1-weighted images and was intermediate between skeletal muscle and fat on T_2-weighted images. The second lesion showed central signal intensity approximating skeletal muscle on T_1-weighted images and fat on T_2-weighted images; the periphery of the tumor was hypointense to skeletal muscle on both pulse sequences [9].

The previously described variable MRI appearance of fibromatosis probably reflects the relative proportion and distribution of collagen, spindle cells, and mucopolysaccharide throughout the tumor. Those lesions hypointense on spin echo MR images are probably markedly hypocellular and contain a large amount of collagen. Conversely, those with high signal intensity are probably quite hypercellular. The corresponding linear and curvilinear regions of decreased signal seen in 86% of the lesions in this series probably reflect areas of dense collagen within the tumor. Awareness of this pattern allowed fibromatosis to be suggested as a preoperative diagnosis in the last patient in this series.

Increased signal on T_1-weighted pulse sequences has been previously reported in lipomas, hematomas, and hemorrhagic neoplasms [11], although not specifically in desmoid tumors. The slightly increased signal intensity relative to skeletal muscle in this series is much less than that seen with lipomas and hemorrhages and may reflect the prolonged repetition time used in these sequences (600–700 ms). Totty et al. published a relatively T_1-weighted (SE900/30) image of a desmoid tumor of the paraspinal muscles that demonstrated a tumor with infiltrating margins [13]. The signal intensity of that tumor was slightly greater than adjacent skeletal muscle, perhaps as a result of the imaging parameters, specifically the added input of spin density to the overall signal intensity in spin echo sequences with long repetition times.

In summary, the MRI appearance of fibromatosis varies. At one extreme the mass is well-defined and "black" on all SE images. At the other, the mass has a nonspecific MRI appearance with long T_1 and T_2 relaxation times (hypointense to skeletal muscle on T_1 and hyperintense to fat on T_2). In our experience, fibromatosis is more commonly a round or oval inhomogeneous mass with well-defined margins and signal intensity between skeletal muscle and subcutaneous fat. Signal voids within the tumor probably represent areas that are relatively hypocellular and with abundant collagen content. Because of the increasingly widespread use of MRI, all radiologists involved in the imaging of soft-tissue masses should be familiar with the MRI appearances of fibromatosis.

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