OBJECTIVE. We present the MRI features with pathologic correlation of aggressive fibromatosis, incorporating 203 cases over a 5-year period from the Royal Marsden Hospital Sarcoma Unit database.

MATERIALS AND METHODS. Sixty patients had imaging available for retrospective review of which 29 had preoperative MRI and final histopathologic diagnosis of aggressive fibromatosis.

RESULTS. The average age at diagnosis was 41.3 years with a female-to-male sex ratio of 1.2:1. Twenty lesions were extraabdominal; six, intraabdominal; and three, in the abdominal wall (classic desmoid). The average tumor size was 6.4 cm (range, 2.2–13.7 cm). Intraabdominal aggressive fibromatosis produced the largest tumors, averaging 9.5 cm. Most lesions were ovoid (52%) or infiltrative (34.5%) in outline with an irregular or lobulated contour (76%). The lesions crossed major fascial boundaries in 31% of cases overall and in 66% of patients referred for recurrent disease. On MRI, homogeneous isointensity or mild hyperintensity on T1-weighted images and heterogeneous high signal on T2-weighted or STIR images were seen. All lesions enhanced after IV gadolinium, usually avidly. In contrast to previous reports, 38% of cases failed to show low signal on all pulse sequences and no abnormalities were seen in local bone structures. Histology showed sheets of bland spindle cells in dense collagen and did not vary with the MRI signal characteristics of the lesion. Patients referred for recurrent disease were most likely to have a recurrence after surgery. MRI and pathology findings did not predict recurrence.

CONCLUSION. Accurate diagnosis and staging of aggressive fibromatosis by MRI have important treatment and prognostic implications.

Fibromatosis is a rare mesenchymal tumor characterized histologically by proliferation of fibroblasts and myofibroblasts with marked production of intercellular collagen. It comprises a broad group of fibrous tissue proliferations of similar histologic appearance that has biologic behavior intermediate between that of benign fibrous lesions and fibrosarcoma [1]. Unlike fibrosarcoma, fibromatosis never metastasizes and is, therefore, classified as a benign neoplasm. However, it is characterized by an infiltrative growth pattern and a tendency toward local recurrence, which have led some to classify it with malignant soft-tissue neoplasms of which it comprises fewer than 4% [2]. The incidence is estimated to be three or four cases per million per year, and it tends to have a female predominance. The peak age of onset has been reported to be between 25 and 35 years, while the majority of cases occur between puberty and 40 years [3]. Most cases are sporadic, but there is a clear association with familial adenomatous polyposis and Gardner’s syndrome, suggesting a link with mutations of the APC gene on chromosome 5q22 [4].

Fibromatosis can be divided into two major groups with several subdivisions by clinical presentation, patient age, and natural history (Appendix 1). Occurring in almost any anatomic location, it can be divided into superficial and deep subtypes. The superficial fibromatoses are usually small (< 5 cm) slow-growing lesions that rarely involve deep structures. The deep group consists of rapidly growing lesions that often reach a large size and have a high tendency to recur after treatment, hence the term “aggressive fibromatosis.” This group principally involves the musculature of the trunk and the extremities. The term “desmoid tumor” (desmos in Greek means band), first used by Mueller in 1838 to emphasize the bandlike or tendonlike consistency of these le-
sions, is synonymous with this type of fibromatosis [1]. However, for this study, we have defined the term “desmoid” to be synonymous with abdominal wall aggressive fibromatosis.

We reviewed the clinicoradiologic features of aggressive fibromatosis from the Royal Marsden Hospital Sarcoma Unit, which has one of the largest databases of soft-tissue tumors worldwide. We assessed multiple MRI parameters to characterize this condition and correlated each case with pathology and risk of postoperative recurrence. Our results are compared with previously published data on aggressive fibromatosis, and we discuss any relevant discrepancies.

Materials and Methods

Permission from the Royal Marsden Hospital Ethics Committee was obtained. Two hundred three patients with biopsy-proven aggressive fibromatosis presented to the Royal Marsden Hospital Sarcoma Unit between 1998 and 2003. Sixty patients had complete imaging records for analysis. Forty-two of those 60 patients underwent CT, of whom 23 had CT as the sole preoperative investigation. Nine patients had sonography, and two patients presenting with a breast mass had mammography. Although 203 patients were referred to the sarcoma unit, only those who had preoperative MRI and available histology were included, of which there were 29 patients in total.

Patient age at presentation and patient sex, tumor type (i.e., primary or recurrent), treatment, and outcome were obtained from the hospital electronic patient record system. The method of diagnosis was recorded—that is, percutaneous biopsy, incisional biopsy, or excisional biopsy. Tumor site, size, shape, contour, and margins; bone changes; crossing of fascial boundaries; and T1, T2, and STIR signal intensities and heterogeneity were assessed on MRI. Tu- mor size was recorded from the MR images as a mean of three orthogonal measurements. Two dedicated sarcoma radiologists reviewed each MRI with findings reached by consensus. The degree of low signal on all sequences, signal intensity from local bone structures, and patterns of behavior after administration of gadolinium were documented. The group was analyzed as a whole and divided into the subgroups previously described in Appendix 1.

Pathology was analyzed for cellularity, pleomorphism, clearance margins, vascularity, and presence of mitoses.

Results

Study Group

Twenty-nine patients with biopsy-proven aggressive fibromatosis had preoperative MRI for analysis. Six patients were referred for recurrent disease. Three of the patients referred for treatment of primary disease underwent surgical excision and had disease recurrence within the time of the study (5 years).

Of the 29 cases, 20 were extraabdominal; six, intraabdominal; and three, in the abdominal wall (desmoid). The average age was 41.3 years with a female-to-male ratio of 1.2:1. Table 1 summarizes the patient demographic data. It should be noted that the desmoid group is slightly younger on average and has a high female predominance (Fig. 1).

The extraabdominal group included the limb girdle and extremity cases. Eleven tumors were located about the shoulder girdle (Fig. 2), five around the pelvic girdle or thigh, two on the trunk (Fig. 3), and two in the popliteal fossa. The intraabdominal group included one case of Gardner’s syndrome (colonic polyposis, osteomas, and abdominal fibromatosis) (Fig. 4).

MRI Features

Twenty-two of the 29 cases had preoperative histologic diagnosis by non-imaging-guided percutaneous core biopsy. One patient underwent sonographically guided core biopsy of a lesion in the root of the neck. Five patients had excisional biopsies, and one patient had an incisional biopsy at another center. The mean tumor size overall was 6.4 cm (range, 2.2–13.7 cm). The intraabdominal group had the largest tumors averaging 9.5 cm (range, 4.3–13.7 cm) in diameter (Fig. 5). Most tumors were either ovoid (52%) (Fig. 1) or infiltrative (34.5%) (Fig. 2) in shape. Seventy-six percent of the cases either had a lobulated or irregular contour (Fig. 3). Fifty-four percent had a well-defined margin, whereas the remainder were either ill defined or partially ill defined (Fig. 4). The adjacent fascia was crossed in nine cases (31%), a finding seen in four (67%) of the six cases referred for recurrent disease (Fig. 3).

All 29 patients had at least one T1-weighted sequence, and 24 cases (83%) were either isointense or slightly hyperintense to skeletal muscle (Fig. 5B). The T1 signal intensity was either homogeneous or mostly homogeneous in 23 cases (79%). On T2-weighted images, 20 (77%) of 26 cases were hyperintense to skeletal muscle (Fig. 1A), of which 17 (65%) returned heterogeneous signal and seven (27%) returned a mostly homogeneous signal.

Moderate to marked low signal across all sequences was identified in nine (31%) of the 29 cases (Figs. 3 and 4). Another nine cases displayed only minimal low signal, and 11 cases (38%) showed no low signal.

After injection of IV gadolinium, all tumors showed enhancement that was either avid (15 cases) (Figs. 5 and 6B) or moderate (three cases) (Fig. 1B) in intensity.

Bone marrow and periosteal signal was normal in every case, despite the close proximity of several of these tumors to bone (Fig. 2). Ten lesions either abutted bone directly or were located within 1 cm of bone. Six lesions were within 2 cm, and no lesion was farther than 4 cm from bone cortex.

Histologic analysis showed sheets of bland spindle cells in dense collagen. Small slitlike vessels were seen, usually with mild perivascular edema and chronic inflammatory cells, especially mast cells. Nuclear atypia was not seen, and few or no mitoses were present (Fig. 7) in all cases. The histology was uniformly the same across the patient group with no differences in primary or recurrent tumors, site, patient sex or age, or MRI signal intensities or heterogeneity.

Treatment and Outcome

Seventeen patients were treated surgically, two of whom had adjuvant chemotherapy or radiation therapy. Eleven patients were treated conservatively. One patient had chemomodulation but no surgery.

Outcome was influenced predominantly by whether the patient was referred for primary

### TABLE I: Demographics of Study Group

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>All Cases</th>
<th>Abdominal Wall (Desmoid)</th>
<th>Intraabdominal</th>
<th>Extrasacral</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of cases</td>
<td>29</td>
<td>3</td>
<td>6</td>
<td>20</td>
</tr>
<tr>
<td>No. of cases that were in recurrence at presentation</td>
<td>6</td>
<td>0</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Age (yr) Mean</td>
<td>41.3</td>
<td>34.3</td>
<td>38.2</td>
<td>44.75</td>
</tr>
<tr>
<td>Range 8–85</td>
<td>30–41</td>
<td>24–54</td>
<td>8–85</td>
<td></td>
</tr>
<tr>
<td>Sex ratio (female: male)</td>
<td>16:13&lt;sup&gt;a&lt;/sup&gt;</td>
<td>3.0</td>
<td>3.3</td>
<td>10:10</td>
</tr>
</tbody>
</table>

<sup>a</sup>1:2:1.
treatment or recurrence. Of the 23 cases referred with primary disease, 13 were treated surgically, nine conservatively, and one received adjuvant therapy only. Of the 13 surgically treated patients, three had postoperative recurrence. In the conservatively managed group, seven were stable, one had a decrease in size, and one had disappeared. The one patient treated with adjuvant therapy only—that is, radiation therapy and chemotherapy—had stable disease 6 years after treatment. Of the six recurrent cases, four had surgery and two had adjuvant therapy. Five had progressive disease, and one was lost to follow-up.

**Discussion**

Aggressive fibromatosis is an important condition that radiologists should include in their differential diagnosis of a soft-tissue mass. On conventional radiographs, aggressive fibromatosis either is undetectable or is identified as an amorphous soft-tissue-density mass (Fig. 6A). Calcification is uncommon. Sonography typically shows a slightly hypoechoic, lobulated, or infiltrative mass. CT will usually show a nonspecific, enhancing soft-tissue mass that may be iso- or slightly hypodense to surrounding muscle (Figs. 4A and 5A).

The principal role of imaging in the management of aggressive fibromatosis is preoperative planning and the detection of recurrence or disease progression in nonsurgically managed patients.

MRI provides the optimum method of tumor evaluation, both before and after surgery because of its superior soft-tissue resolution compared with other imaging techniques. The relationship of the tumor to important local structures, such as nerves, vessels, deep viscera, and bone, can only be accurately assessed using MRI. Involvement of these structures is likely to influence initial patient management and may represent tumor recurrence if seen on follow-up scans.

The main clinical differential diagnosis of aggressive fibromatosis is malignant soft-tissue sarcoma, extranodal lymphoma, and benign myositis ossificans and arteriovenous malformation. The two benign diagnoses can be characterized on imaging by showing typical calcification on conventional radiography or CT in the former and signal flow voids and calcification on MRI in the latter. Extranodal lymphoma presenting as a muscular mass is rare.

Therefore, the main differential diagnosis of aggressive fibromatosis on imaging is malignant soft-tissue sarcoma. Can MRI accurately differentiate aggressive fibromatosis from soft-
tissue sarcoma? As a general rule, most soft-tissue sarcomas grow as space-occupying intramuscular lesions, enlarging in a centripetal fashion that pushes rather than infiltrates local structures. They typically form a pseudocapsule as they enlarge by compressing normal tissue and usually respect fascial boundaries, remaining within anatomic compartments until late in their course. As they enlarge further, soft-tissue sarcomas may outstrip their blood supply resulting in central necrosis [5]. In contrast, our series reveals that aggressive fibromatosis typically shows an infiltrative growth pattern; crosses fascial boundaries; and does not show central necrosis, even in the largest lesions. Additional MRI findings that point toward the diagnosis of aggressive fibromatosis are bands of low signal intensity across all se-

Fig. 2—28-year-old woman who presented with painful lump in upper arm. Axial T1-weighted MR image obtained after injection of IV gadolinium shows mass in left deltoid muscle. Bone marrow and periosteal signal are normal. Note infiltrative pattern of spread into subcutaneous tissues (black arrows) and tumor crossing fascial boundaries into flexor (arrowhead) and extensor (white arrow) compartments of arm. This is typical pattern in aggressive fibromatosis. In this case, extensive local spread deemed this lesion to be inoperable and patient was treated with adjuvant therapy only. Lesion has been stable for 36 months.

Fig. 3—22-year-old woman who presented with multicentric fibromatosis since childhood. A and B, Axial (A) and coronal (B) T2-weighted MR images through trunk show large, lobulated, mixed-signal-intensity mass (arrows, A) arising within erector spinae muscles affecting both sides of midline. Note extensive area of low signal (asterisks), typical of aggressive fibromatosis.
sequences and uniform moderate to avid enhancement after gadolinium administration. Low signal intensity on all pulse sequences is said to be characteristic of, but not specific for, aggressive fibromatosis [6]. Initial reports emphasized this finding and used it as a means of differentiating aggressive fibromatosis from malignant soft-tissue sarcomas [7]. However, low signal is not seen in all cases of aggressive fibromatosis, and prominent low signal intensity, particularly on T2-weighted images, may be seen in pigmented villous nodular synovitis, giant cell tumors of the tendon sheath, calcified masses, and malignancies such as fibrosarcoma or malignant fibrous histiocytoma [6]. These lesions complete the differential diagnosis.

It should be stressed that although MRI can provide clues to the correct diagnosis of aggressive fibromatosis, tissue confirmation is required for a final diagnosis in all cases. Aggressive fibromatosis generally presents as a

Fig. 4—24-year-old man with Gardner's syndrome who presented with abdominal pain. A, Axial contrast-enhanced CT image of abdomen shows amorphous soft-tissue mass (arrowheads) in mesentery. Note presence of stent (arrow) in pelvis of right kidney. Fibromatosis extended inferiorly into pelvis obstructing right ureter. B, Axial T2-weighted MR image through abdomen shows mixed-signal-intensity mass (arrowheads) with prominent areas of low signal (arrows), believed to represent fibrosis. Percutaneous biopsy confirmed diagnosis of intraabdominal fibromatosis.

Fig. 5—52-year-old man who presented with abdominal mass. A, Axial contrast-enhanced CT image of abdomen shows uniformly dense soft-tissue mass (arrows) in small-bowel mesentery. B, Axial unenhanced T1-weighted MR image through abdomen shows large mostly homogeneous, well-defined, ovoid mass lesion in peritoneal cavity (arrows). Mass was fully resected confirming diagnosis as intraabdominal fibromatosis. No recurrence was seen after 2 years of follow-up.
palpable abnormality within the peripheral musculature, which is usually amenable to percutaneous biopsy. The majority of the patients in this series (22/29) underwent non-imaging-guided percutaneous core biopsy in the surgical outpatient clinic and avoided an invasive diagnostic procedure under anesthetic. For more complex masses involving the abdominal wall or peritoneum, imaging guidance may be required. In all cases, at least three 14-gauge cores should be obtained for accurate histologic assessment [8].

In our experience, the most common findings of aggressive fibromatosis on MRI are of an ovoid or infiltrative lesion that is lobulated or irregular in outline and crosses fascial boundaries in almost one in three cases. An infiltrative growth pattern is seen almost twice as often in the recurrent cases than in the primary cases, which may reflect the more aggressive nature of these lesions. Typically, aggressive fibromatosis lesions are homogeneously isointense on T1-weighted images, are heterogeneous high signal on T2-weighted and STIR images, and enhance avidly after IV gadolinium administration. Bands of low signal intensity within the lesion on all sequences were seen in 18 (62%) of the 29 cases. These findings are generally in concordance with previously published data [5, 6, 9–13].

Where this study differs from previously published work on aggressive fibromatosis is the absence of changes in local bone structures and in the degree of low signal within the lesion. Previous studies of aggressive fibromatosis suggest that bone changes occur in up to 37% of the cases [14], a finding not backed up by our series. It is recognized that bone changes are more common in recurrent aggressive fibromatosis [15], although this was not seen in the three extraabdominal recurrent cases in our study. No abnormalities of local bone structures were seen on MRI despite 20 of the cases being extraabdominal in location, with no lesion being farther than 4 cm from bone. The discor-

Fig. 6—60-year-old man who presented with incidental finding on conventional radiography.
A, Conventional radiograph shows clearly marginated right apical soft-tissue noncalcified mass (arrowheads) that has appearance of Pancoast’s tumor. B, Contrast-enhanced T1-weighted coronal MR image shows enhancing mass (arrowheads) in right apex. Despite mimicking Pancoast’s tumor, repeated biopsies confirmed diagnosis as fibromatosis.

Fig. 7—31-year-old man who presented with mass in thigh. Photomicrograph of H and E–stained histopathology slide shows typical features of fibromatosis. Note bland sheets of spindle cells in dense collagen stroma. Slitlike vessel is seen with minor perivascular edema and chronic inflammatory cells including lymphocytes and mast cells. Note also absence of mitoses or nuclear atypia. This appearance is seen in both primary and recurrent cases.
dance in bone changes may be explained by the small numbers of cases involved in our study and previous studies, which vary from three [16] to 40 [9]. In addition, most of the previous descriptions are based on radiography findings rather than MRI findings [14].

Although bands of low signal intensity were seen in all sequences in 62% of the cases of this series, this finding was classified as marked in only three cases (10%). Furthermore, this finding was classified as minimal or absent in 20 (69%) of the 29 cases in our study. It has been hypothesized that the lack of low signal in some cases of aggressive fibromatosis may be due to the cellular component within these lesions. Sundaram et al. [16] attempted to explain this variability by comparing the T2-weighted MRI appearances with pathology in three cases of aggressive fibromatosis. Two of these lesions had decreased signal intensity on T2-weighted images, which was said to reflect hypocellularity and abundant collagen on histology. In the third case, high signal intensity on T2-weighted images was seen, and this correlated with marked cellularity [16]. In our study, as in larger subsequent studies, the MR–pathologic findings of aggressive fibromatosis are more variable [9, 10]. The histopathologic findings in our series were uniform across all cases, independent of signal characteristics on MRI. The heterogeneous T2 and STIR signal intensity pattern, which was the most common pattern in our study, may correspond to varying proportions of cellular tissue, myxoid tissue, and collagen in each lesion [9], although this could not be accurately quantified in our samples.

Compared with other forms of aggressive fibromatosis, abdominal wall desmoid tumors are known to have a lower postoperative recurrence rate and to occur in younger women, frequently around the time of pregnancy [17]. Only three cases were included in this study, making meaningful analysis of this subgroup difficult. The small number in this series presumably reflects common surgical practice in the United Kingdom, where these lesions are resected in secondary rather than tertiary centers. The risk of recurrence was not related to patient age or sex or to tumor site or size. On MRI, tumor shape, contour, margins, T1 and T2 signal intensities, and contrast enhancement patterns did not predict recurrence but did help to characterize the lesion. As in other rare conditions, the relatively small numbers involved made statistical analysis difficult. The only major risk factor for postoperative recurrence was if the patient was referred for recurrent disease. In five of the six patients referred for recurrent disease in our study, disease recurred or progressed during the study interval.

Management of extraabdominal fibromatosis is a challenge because this disease does not respect the usual surgical rules relating to resection and recurrence. Surgical excision with clear margins is the goal. However, if the tumor is deemed inoperable because of involvement of significant neurovascular structures or poor patient performance status, then conservative management, adjutant treatment with radiation therapy [18], chemotherapy, or hormonal antiestrogen treatment can be used [19]. It is important to note that after a period of rapid and alarming growth of either primary or recurrent disease, spontaneous growth arrest may take place, the so-called “plateau phase,” and can be maintained indefinitely [20]. Results of treatment need to reflect this because growth arrest cannot be claimed as a therapeutic success. The most convincing supporting data come from the Mayo Clinic where stable disease was seen in 60 of 68 patients over an average follow-up period of 6.3 years [21].

In conclusion, aggressive fibromatosis is the likely diagnosis if, on MRI, the mass is ovoid or irregular, crosses fascial compartments, is isointense on T1- and heterogeneously hyperintense on T2-weighted images, and shows bands of low signal on all sequences. Local changes in bone are not typical. A core-cut biopsy, which may be imaging-guided, is sufficient for histopathologic characterization. MRI is the imaging method of choice for both preoperative planning and posttreatment monitoring. Aggressive fibromatosis is a challenging disease for both surgeons and radiologists alike. Awareness of this diagnosis in a young or middle-aged adult presenting with a mass, particularly around a limb girdle or in the abdominal wall of a woman, should enable optimum therapy to be instituted.

References

Appendix I appears on the next page
APPENDIX 1: Classification of Fibromatosis (Adapted from [1])

Superficial (fascial) fibromatosis
- Palmar fibromatosis (Dupuytren’s disease)
- Plantar fibromatosis (Ledderhose’s disease)
- Knuckle pads

Deep (musculoaponeurotic) fibromatosis
- Extraabdominal fibromatosis (extraabdominal desmoid tumor)
- Abdominal wall fibromatosis (abdominal desmoid tumor)
- Intraabdominal fibromatosis (intraabdominal desmoid tumor)
  - Pelvic fibromatosis
  - Mesenteric fibromatosis
  - Mesenteric fibromatosis in Gardner’s syndrome