

MRI and Biologic Behavior of Desmoid Tumors in Children

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OBJECTIVE. The outcome of desmoid tumor in children cannot be reliably predicted on the basis of histologic findings. We sought to determine whether the postoperative presence of residual or recurrent tumor can be predicted on the basis of demographic variables and baseline MRI features of the tumor. We also aimed to determine how imaging features change during adjuvant treatment and how the imaging features relate to the histologic features.

MATERIALS AND METHODS. Two radiologists retrospectively reviewed images from 281 MRI examinations performed at baseline and during postoperative therapy for desmoid tumor. The examinations had been performed on 17 children treated between September 1991 and March 2003. Tumor volume; distinctness of margins; involvement of bone and neurovascular bundle; and T1-weighted, T2-weighted, and STIR signal intensity and contrast enhancement pattern were recorded. Baseline imaging and demographic features were correlated with the postoperative presence of residual or recurrent tumor. Imaging changes during follow-up were compared with treatment response and outcome. The imaging features of eight tumors were compared with percentage cellularity and collagen deposition in biopsy samples obtained within 30 days of imaging.

RESULTS. Baseline involvement of the neurovascular bundle approached significance as a predictor of the presence of residual or recurrent tumor ($p = 0.08$). Other baseline imaging and demographic features were not predictive ($p \geq 0.4$). Changes in imaging features were variable during follow-up. T2-weighted and STIR signal intensity may be correlated with percentage cellularity and collagen deposition.

CONCLUSION. MRI has limited value in prediction of the postoperative presence of residual or recurrent desmoid tumor in children. It is useful, however, for detecting disease and monitoring postoperative adjuvant therapy.

Fibromatosis of childhood is a broad spectrum of fibrotic neoplasms that include congenital fibromatosis, congenital fibrosarcoma, fibromatosis coli, and desmoid tumor (also known as aggressive fibromatosis or desmoid-type fibromatosis). The most common of these conditions is desmoid tumor, a monoclonal fibroblastic proliferation arising in musculoaponeurotic tissues. Desmoid tumor is classified by the World Health Organization as a tumor of intermediate grade. Although desmoid tumor does not metastasize and associated mortality is rare, loss of muscle and joint function is often associated with tumors that develop in the extremities. The optimal treatment is controversial; a multidisciplinary approach including surgery, chemotherapy, and radiation therapy is often used. The goal of treatment is to provide local control while preserving function and appear-

ance. Surgery historically has been the primary treatment. However, achieving the wide margin that offers the best chance of avoiding recurrence is difficult because of the infiltrative nature of desmoid tumors, and local recurrence after surgical excision is common [1, 2]. Postoperative adjuvant therapies have included radiation therapy and cytotoxic and noncytotoxic chemotherapy (e.g., nonsteroidal antiinflammatory drugs and estrogen antagonists), but the appropriate use of these treatments in the pediatric population needs to be defined. Of particular concern in children are the long-term effects of such treatments in the management of a relatively benign disease [2, 3]. Because the disease is rather rare, few reports have described the imaging features, treatment, and outcome among pediatric patients [2, 4]. Therefore, we reviewed our experience with desmoid tumors in children to determine whether demo-

graphic variables and MRI findings are associated with tumor recurrence, histologic features, treatment response, and outcome.

Materials and Methods

Imaging, Medical Record, and Pathologic Review

Between September 1991 and March 2003, 17 children were referred to our institution for evaluation of desmoid tumor, seven for recurrent or residual disease after treatment at a local institution. After obtaining institutional review board approval, we reviewed the MR images and medical records. Our review was conducted in compliance with the Health Information Portability and Accountability Act of 1996. Joint review of baseline (before intervention) and follow-up (after intervention) images was performed by two pediatric radiologists who were aware of the diagnosis of desmoid tumor but did not know the patient's age, treatment history, or disease outcome. For each imaging examination, the reviewers recorded the number and anatomic location of the tumors and the volume of each tumor using the following formula: (anteroposterior diameter × transverse diameter × length) × 0.52.

Baseline tumor images were assessed for local tumor infiltration by determination of the percentage of tumor margin that was indistinct (not sharply defined from surrounding soft tissue) on the sequence that best showed the tumor margins. Baseline images also were assessed for involvement of the neurovascular bundle and bone, defined as abutting if tumor touched the structure, encasing if tumor surrounded any part of it, and direct invasion if tumor invaded it. Baseline T1-weighted images were assessed to determine the predominant (≥ 50%) tumor signal intensity, defined as isointense, hypointense, or hyperintense relative to adjacent normal muscle.

Because previous reports [5, 6] have suggested that areas of desmoid tumor that are hyperintense on T2-weighted and STIR images are associated with active fibroblastic proliferation, baseline and follow-up images were reviewed for subjective determination of the percentage of the entire tumor volume that was hyperintense to normal muscle on these sequences and the predominant signal intensity. If fat had higher signal intensity than muscle on these sequences, the images were considered inadequately fat suppressed, and they were excluded from analysis. The sequence that gave the higher predominant signal intensity was used for analysis when there was a discrepancy. Tumors lacking a predominant signal intensity were considered of mixed intensity. Baseline and follow-up images also were subjectively assessed for percentage of the entire tumor volume that was contrast enhanced and overall enhancement intensity, which was graded as mild (equal in signal intensity to muscle),

moderate (higher signal intensity than muscle but lower signal intensity than blood vessels), or intense (same signal intensity as or higher signal intensity than blood vessels).

For tumors on which follow-up was conducted until the end of therapy, imaging changes that occurred between the imaging date closest to initiation of treatment and either immediately before a change in treatment technique (radiation therapy vs chemotherapy vs surgery) or the most recent follow-up evaluation were reported. Using the criteria established for the Pediatric Oncology Group 9650 and Children's Oncology Group ARST0321 desmoid tumor studies, we assessed tumor response on follow-up images as the product of the largest bidirectional tumor diameters as follows: progressive disease if the product increased more than 25% above the smallest product of bidimensional diameters recorded; stable disease if the product showed a 25% or smaller increase or decrease; partial response if the product decreased more than 50%; and minor response if the product decreased between 25% and 50%. Tumors were considered completely resolved when no tumor was identified on MR images.

From medical records we recorded patient age at diagnosis, sex, medical history, operative and pathologic reports, clinical course, and outcome of desmoid tumor treatment. Because desmoid tumor can be associated with familial adenomatous polyposis, we also recorded family history of familial adenomatous polyposis. Pathologic specimens obtained within 30 days of MRI were reviewed by one pathologist, who graded the amount of collagen deposition and the percentage cellularity of each specimen. Collagen deposition was graded as follows: 1, rare bands; 2, readily notable collagen; 3, abundant collagen. Percentage cellularity was based on the average cellularity seen in 10 random high-power fields.

Statistical Analysis

For statistical analysis, residual and recurrent tumor after surgical resection was considered a single outcome, and patients were categorized as having or not having residual or recurrent tumor. We used the Wilcoxon and Mann-Whitney tests to compare age, baseline percentage tumor enhancement, baseline tumor enhancement intensity, baseline tumor percentage indistinct margin, and baseline tumor volume of patients with recurrent or residual tumors and those without. We analyzed the association between sex, baseline T1-weighted signal intensity, baseline tumor involvement of the neurovascular bundle or bone, and residual or recurrent tumor by using the Fisher's exact test. For the purposes of analysis, baseline tumor involvement of the neurovascular bundle and bone was a dichotomous variable (involvement vs no involvement). In-

volvement was defined as abutting, surrounding, or invading. Tumor enhancement intensity was considered a dichotomous variable: intense versus moderate or mild. No statistical test was performed for baseline tumor T2-weighted or STIR signal intensity because all tumors were predominantly hyperintense on these sequences. The analyses were performed with SAS v9.1 (SAS Institute) and StatXact PROCs v6.2 (Cytel) software.

Results

Demographic Features and Medical History

The study group comprised seven girls and 10 boys with a mean age at diagnosis of 7 years 10 months (range, 2 months to 19 years 10 months). We found no correlation between age at diagnosis ($p = 0.7$) or sex ($p = 0.6$) and disease recurrence (Table 1). One patient had familial adenomatous polyposis (Gardner syndrome) (Fig. 1).

Management and Outcome

One patient was lost to follow-up. The follow-up times of the other 16 patients ranged from 6 months to 9 years 7 months with an

TABLE 1: Correlation Between Baseline Tumor Demographic and MRI Variables and Presence of Residual or Recurrent Desmoid Tumor After Surgical Resection

Demographic Variable	<i>p</i>
Age at diagnosis	0.70
Sex	0.60
Baseline tumor MRI variable	
Tumor volume	0.40
Predominant T1-weighted signal intensity	1.0
Predominant T2-weighted or STIR signal intensity	NA ^a
Percentage of tumor volume enhancement	0.5
Intensity of tumor enhancement	1.0
Percentage of tumor margin indistinct	0.40
Bone involvement	0.50
Neurovascular bundle involvement	0.08
Both bone and neurovascular bundle involvement	NA ^b

^aNA = not statistically evaluated because all evaluable tumors ($n = 5$) were predominantly hyperintense on T2-weighted and STIR images.

^bNA = not statistically analyzed because all patients with tumors that involved both the neurovascular bundle and bone ($n = 4$) also had residual ($n = 3$) or recurrent ($n = 1$) tumor after surgical resection.

MRI of Desmoid Tumors



Fig. 1—19-year-old male with Gardner syndrome and massive desmoid tumor of paraspinal musculature. **A**, Baseline sagittal T2-weighted MR image (TR/TE, 5,950/116) shows hypointense bands (*arrows*) coursing through hyperintense tumor. **B** and **C**, Sagittal unenhanced T1-weighted MR image (618/14, **B**) and contrast-enhanced T1-weighted image (804/14, **C**) show lack of enhancement of hypointense bands (*arrows*) in **A**. These nonenhancing, hypointense bands are probably areas of fibrosis within enhancing active fibroblasts. Patients with Gardner syndrome or familial adenomatous polyposis have 20% lifetime risk of extraabdominal or, more commonly, intraabdominal desmoid tumor.

average follow-up time of 4 years 5 months. Sixteen of the 17 patients received treatment with up-front surgical resection. Four of these patients had no residual or recurrent tumor after surgery, six had residual tumor, and six had recurrent tumor in or near the surgical bed. One patient had an unresectable chest wall tumor, which was treated with up-front radiation therapy and then subtotal surgical resection and chemotherapy. This patient had a partial response. For the 12 patients with residual or recurrent disease, treatment varied considerably. Five received treatment with repeated resection alone. The tumors of two patients were

grossly resected. One of these tumors recurred multiple times (Fig. 2), and the affected finger was amputated to achieve final control. The other patient was lost to follow-up. Tumors of the other three patients were subtotally resected; two residual tumors remained stable, and one progressed, but the patient did not receive additional treatment. The other seven patients with residual or recurrent tumors received combinations of chemotherapy ($n = 5$), surgery ($n = 4$), and radiation therapy ($n = 4$). One of these seven patients had a complete response; two, a minor response; and two, a partial response. Two patients had progressive disease.

Therefore, after surgery alone ($n = 10$) or combined treatment ($n = 7$), five patients had a complete response; two, a minor response; three, a partial response; two, stable disease; four, progressive disease; and one was lost to follow-up.

Imaging Findings

The 17 patients underwent a total of 281 MRI examinations (mean, 17 examinations per patient; range, one to 22). Baseline images of the original tumor, before any intervention, were available for 10 patients. T1-weighted images were available for all 10 patients, T2-weighted or STIR images for five

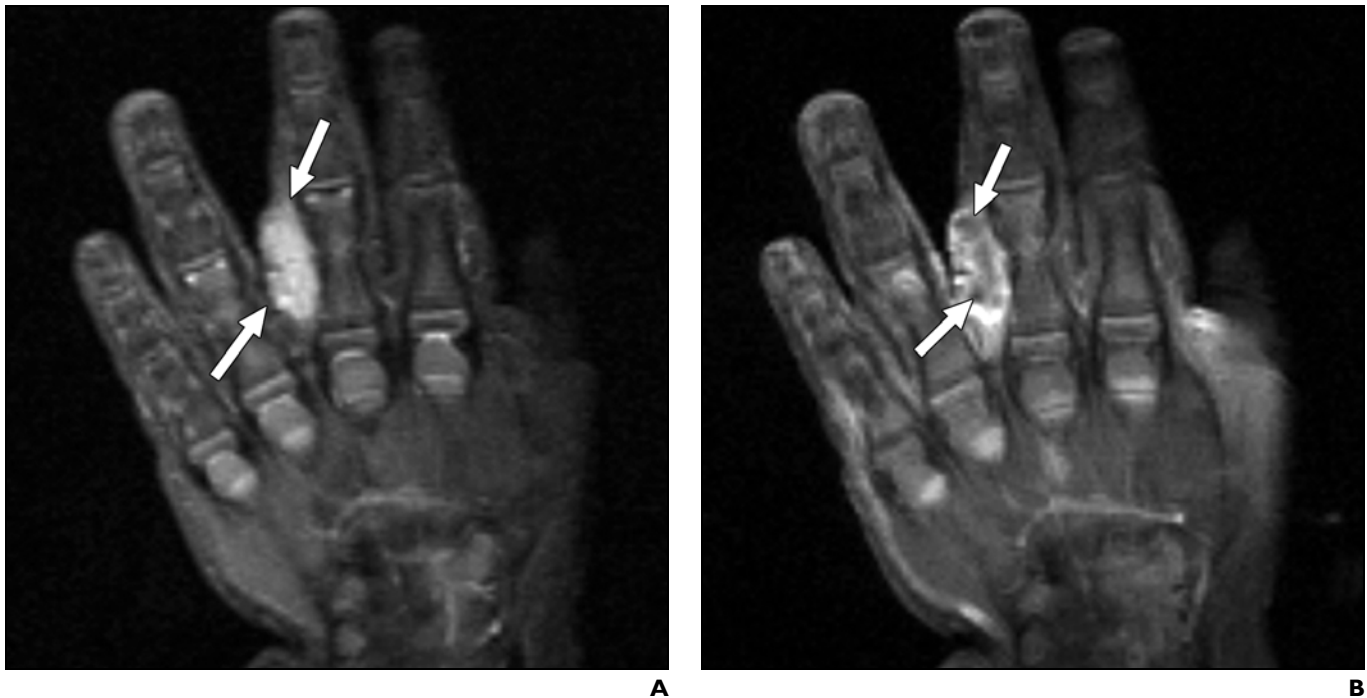


Fig. 2—10-month-old girl with desmoid tumor of middle finger. **A**, T2-weighted coronal image (TR/TE, 3,000/30) shows predominantly hyperintense mass (arrows) with fairly sharp margins, except for proximal and distal margins. **B**, T1 contrast-enhanced coronal image (400/20) shows peripheral tumor enhancement and central lack of enhancement (arrows). Although small, this tumor necessitated amputation of digit for local control.

TABLE 2: Baseline Tumor MRI Features and Assessment of Presence of Residual or Recurrent Tumor After Surgical Resection (n = 10)

Tumor Location	Tumor Volume (mL)	Signal Intensity		Percentage Tumor Volume Enhancement	Intensity of Enhancement	Percentage Tumor Margins Indistinct	Neurovascular Bundle Involvement	Bone Involvement	Residual or Recurrent Tumor
		Predominant T1-Weighted Images ^a	Predominant T2-Weighted or STIR Images ^a						
Paraspinal	1,246	Isointense	Hyperintense	70	Intense	20	No	No	Residual
Chest wall	466	Isointense	Hyperintense	95	Intense	30	Encased	Encased	Residual
Gluteal	234	Isointense	Hyperintense	85	Intense	10	No	No	No
Mediastinal	187	Isointense	NA	NA	NA	20	Abutted	Invaded	Residual
Chest wall	121	Isointense	Hyperintense	50	Mild	30	Abutted	Abutted	Residual
Lower extremity	76	Isointense	NA	90	Intense	20	Abutted	Abutted	Recurrent
Paraspinal	43	Isointense	NA	80	Intense	10	No	Abutted	Recurrent
Upper extremity	26	Isointense	Hyperintense	80	Moderate	20	No	Abutted	No
Chest wall	7	Hypointense	NA	NA	NA	0	No	Abutted	Residual
Lower extremity	3	Isointense	NA	100	Intense	40	No	No	No

Note—NA = images not available or images inadequate for review.
^aSignal intensity relative to normal muscle.

patients, and contrast-enhanced images for eight patients. Imaging features and outcome after surgical resection of these 10 baseline tumors are summarized in Table 2. Three of the five baseline tumors imaged with T2-weighted or STIR sequences had predominantly high signal intensity with hypointense

areas and bands interspersed throughout the tumor. These areas and bands did not become enhanced after contrast administration (Figs. 1 and 2). Such areas were found in many of the recurrent tumors. The correlation between baseline tumor imaging features and outcome is shown in Table 1. Involvement of

the neurovascular bundle approached significance as a predictor of the presence of residual or recurrent tumor ($p = 0.08$). In four patients, baseline tumors involved both the neurovascular bundle and bone, and all four patients had residual ($n = 3$) or recurrent tumor ($n = 1$) after surgical resection. None of

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TABLE 3: Changes in MRI Parameters of Desmoid Tumors Followed Through Radiation Therapy and Chemotherapy

Tumor Number	Change in Tumor Volume	Change in Percentage Tumor Hyperintensity on T2-Weighted or STIR Images	Change in Percentage Tumor Enhancement	Change in Intensity of Enhancement
Tumors followed through radiation therapy				
1	↑	↓	NA	NA
2	↓	↓	↑	↔
3	↓	↓	↓	↔
Tumors followed through chemotherapy				
1	↓	NA	↓	↓
2	↓	↑	↑	↔
3	↓	↑	↔	↔
4	↓	↓	↓	↓
5	↓	↓	↓	↓
6	↓	↔	↔	↔
7	↑	↓	↓	↓
8	↔	↓	↑	↔

Note—↑ = increased over time, ↓ = decreased over time, ↔ = did not change over time, NA = images not available or images inadequate for review.

the other imaging parameters was a significant predictor of the presence of recurrent or residual disease.

Posttreatment MR images were available for 16 patients. Four patients were treated with radiation, one for presumed residual tumor after total gross resection. This patient did not have convincing imaging evidence of residual tumor on follow-up MRI examinations. The changes in tumor imaging features that occurred during radiation therapy in the other three patients are shown in Table 3. One of these tumors increased in size and two became smaller. The percentage of tumor volume that was hyperintense on T2-weighted or STIR images decreased in all three tumors (Fig. 3). In three patients new desmoid tumors developed within or near the radiation field. All of the patients were clinically considered to have progressive disease, and they underwent additional surgical resection or chemotherapy. Six patients (eight desmoid tumors) underwent follow-up until the end of chemotherapy. The evolution of MRI features during chemotherapy was variable from person to person and is summarized in Table 3. Follow-up MRI showed one tumor completely resolved.

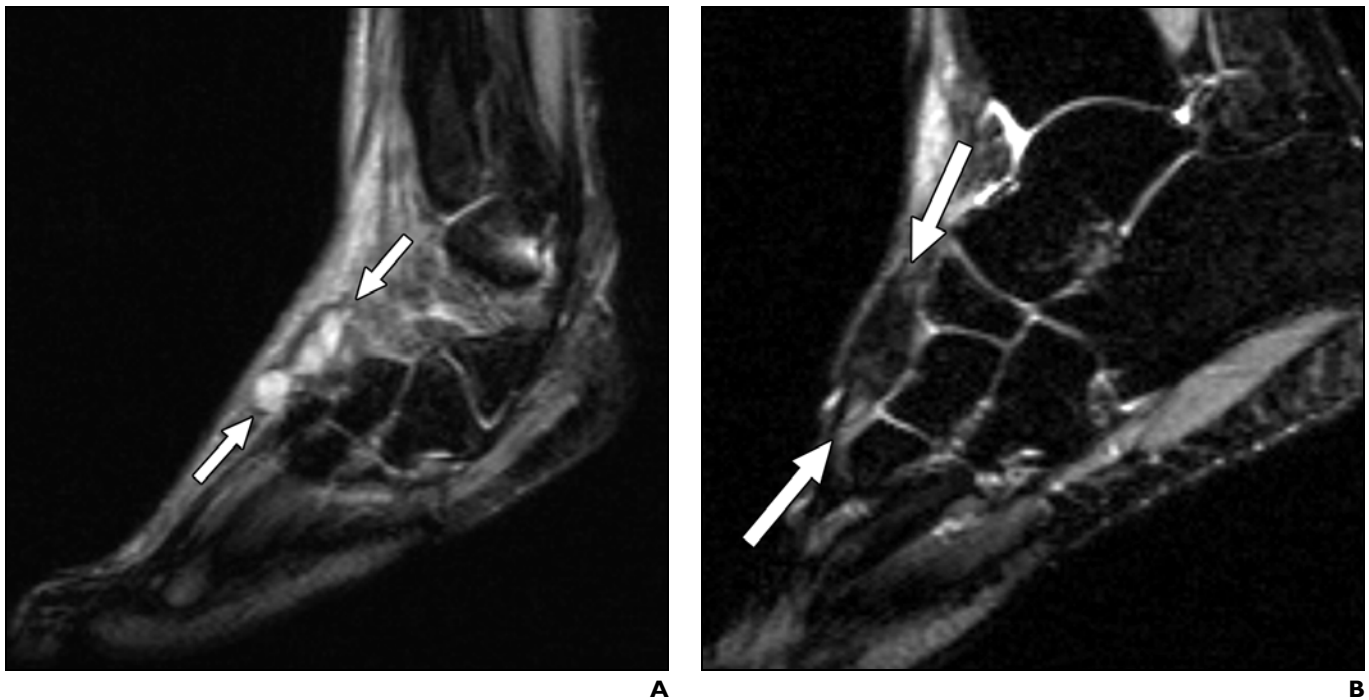


Fig. 3—14-year-old girl with recurrent desmoid tumor in left foot.

A, Sagittal STIR image (TR/TE, 2,500/18; flip angle, 140°) 2 weeks after completion of radiation therapy shows tumor (arrows) predominantly hyperintense to muscle. Some of hyperintense signal may be result of edema from recent radiation.

B, Sagittal STIR image (3,500/18; flip angle, 140°) 2 years 7 months after **A** shows tumor (arrows) has grown slightly but is predominantly hypointense to muscle.

TABLE 4: Histologic and Corresponding MRI Features of Desmoid Tumors Biopsied Within 30 Days of MRI (n = 8)

Histologic Features		Tumor MRI Features		
Percentage Cellularity	Collagen Deposition	Percentage Hyperintensity to Muscle on T2-Weighted or STIR Images	Enhancement	
			Percentage	Intensity
5	3	0	30	Moderate
20	3	NA	100	Intense
20	3	NA	80	Intense
30	2	NA	85	Intense
30	2	NA	90	Intense
40	2	75	85	Intense
40	1	70	70	Intense
50	1	90	90	Moderate

Note—Collagen deposition graded as 1 = rare bands, 2 = readily notable collagen, 3 = abundant collagen, NA = images not available or images inadequate for review.

Histology and Associated MRI Findings

Of 44 resections or biopsy procedures, eight were conducted within 30 days of MRI. Histologic and corresponding MRI findings for these eight are shown in Table 4. The relation between percentage cellularity and collagen deposition was inverse. Tumors with higher percentage cellularity had less collagen deposition than did those with lower percentage cellularity. The tumor with the least cellularity (5%) and abundant collagen had the least percentage volume enhancement (30%) (Fig. 4). The tumor with the greatest percentage cellularity (50%) and rare bands of collagen had enhancement of 90% of the tumor volume (Fig. 5).

Discussion

Desmoid tumor, the most common type of fibromatosis in children, is characterized by intertwining fibroblasts and myofibroblasts arranged in broad bundles that infiltrate

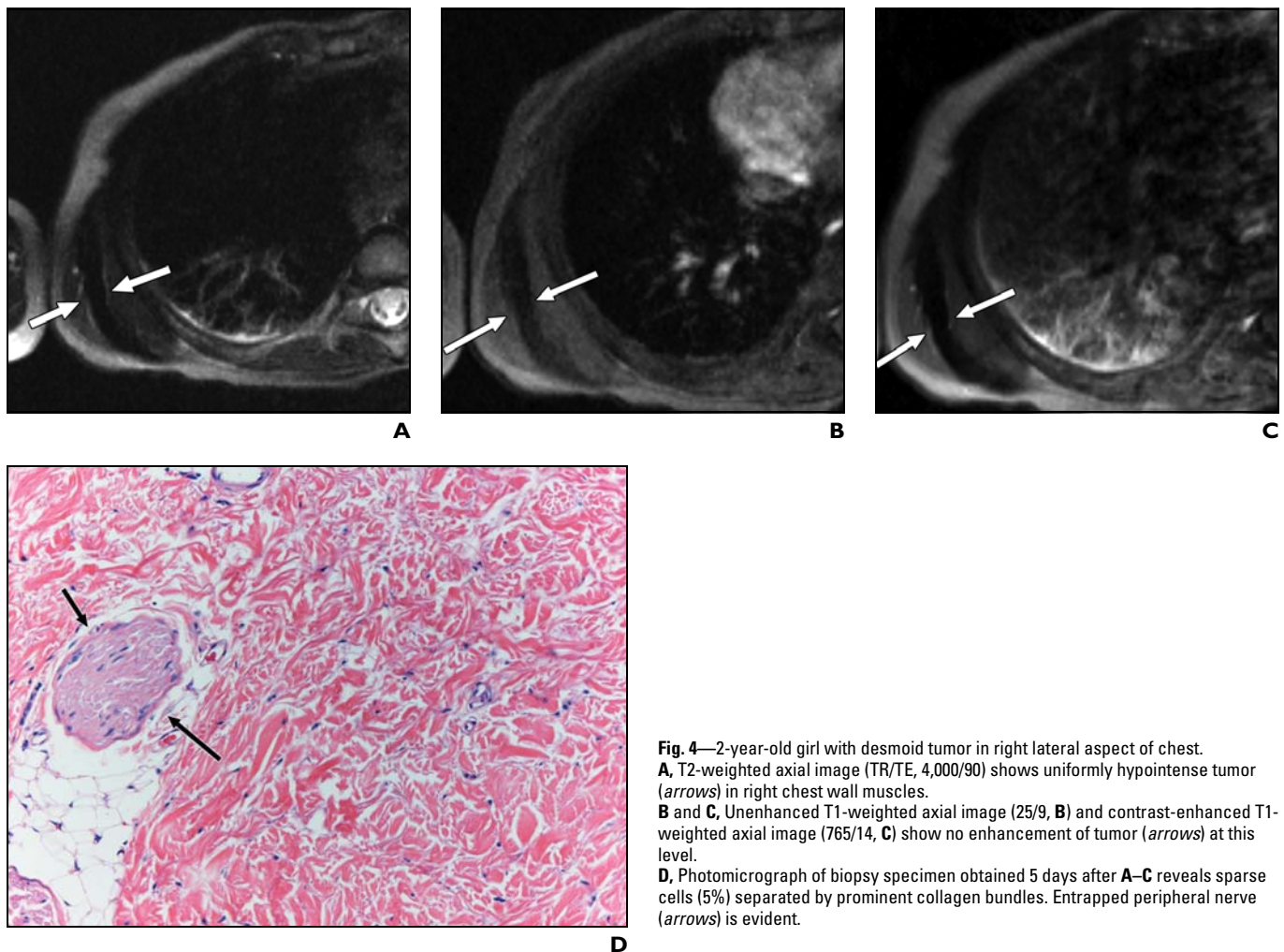


Fig. 4—2-year-old girl with desmoid tumor in right lateral aspect of chest. **A**, T2-weighted axial image (TR/TE, 4,000/90) shows uniformly hypointense tumor (arrows) in right chest wall muscles. **B** and **C**, Unenhanced T1-weighted axial image (25/9, **B**) and contrast-enhanced T1-weighted axial image (765/14, **C**) show no enhancement of tumor (arrows) at this level. **D**, Photomicrograph of biopsy specimen obtained 5 days after **A–C** reveals sparse cells (5%) separated by prominent collagen bundles. Entrapped peripheral nerve (arrows) is evident.

MRI of Desmoid Tumors

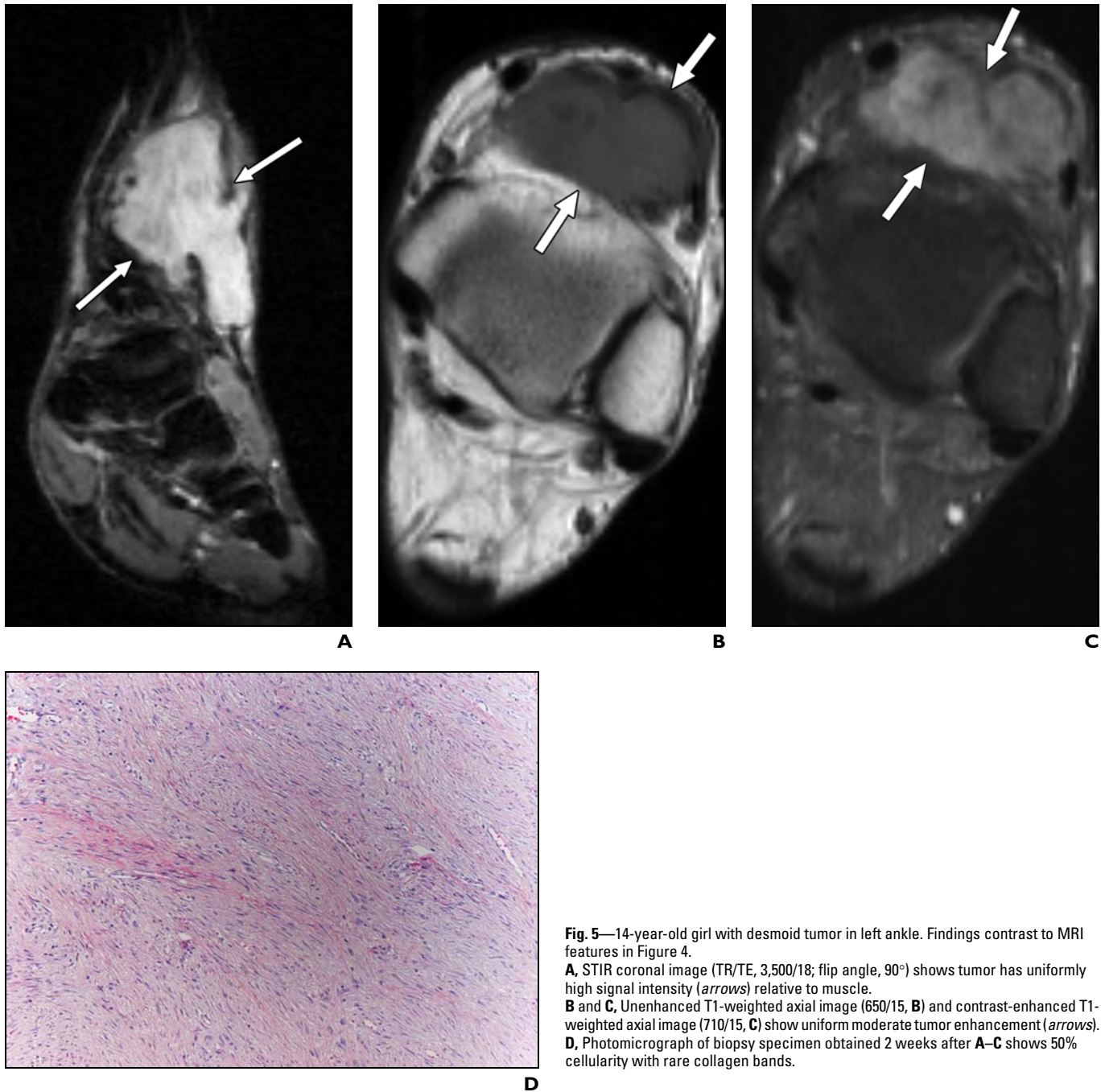


Fig. 5—14-year-old girl with desmoid tumor in left ankle. Findings contrast to MRI features in Figure 4.

A, STIR coronal image (TR/TE, 3,500/18; flip angle, 90°) shows tumor has uniformly high signal intensity (*arrows*) relative to muscle.
B and C, Unenhanced T1-weighted axial image (650/15, **B**) and contrast-enhanced T1-weighted axial image (710/15, **C**) show uniform moderate tumor enhancement (*arrows*).
D, Photomicrograph of biopsy specimen obtained 2 weeks after **A–C** shows 50% cellularity with rare collagen bands.

surrounding structures and are usually surrounded by abundant collagen. Although benign, desmoid tumors have variable biologic potential, which cannot be predicted on the basis of histologic features, and tend to recur after local excision. The nomenclature used to describe these tumors in children is confusing. Terms such as infantile and juvenile fibromatosis imply distinctive biologic properties in different age groups [7–11]. Such distinctive fea-

tures are not evident in the published literature, and although our cohort was small, we found no difference in the clinical behavior or MRI features of these tumors among patients of different ages. In our study, nine of 10 primary tumors were isointense in relation to muscle on T1-weighted images, whereas T2-weighted and STIR images showed predominantly high signal intensity. Contrast enhancement was intense in most of the primary tumors. Most of

the tumors had enhancement of 80% or more of the tumor volume. Tumor margins ranged from 100% sharply defined to 40% infiltrative. None of these MRI characteristics was predictive of the presence of residual or recurrent disease.

Thirteen (77%) of our patients had residual or recurrent tumor after up-front surgical resection. We found that patients with tumors involving the adjacent neurovascular bundle or both the neurovascular bundle and bone were more

likely than others to have residual or recurrent tumor after surgical resection. This finding is not surprising because tumors involving these structures are difficult to resect completely without morbidity. Our finding is consistent with the results of a 1995 retrospective study [12] of pediatric patients with desmoid tumors that showed surgical resection with negative margins was associated with a 70% recurrence-free survival rate. Therefore, we favor surgical treatment of children with desmoid tumor if the operation is likely to result in complete resection and if it can be performed without undue morbidity, such as amputation. In our cohort, patients with residual or recurrent disease who were treated with chemotherapy fared better than those treated with radiation therapy. One of six patients treated with chemotherapy had progressive disease, compared with three of four patients treated with radiation. An interesting finding was that additional tumors developed within or near the radiation field of three of the four patients treated with radiation therapy, although a causal relation was not established because of the small sample size.

Although our sample size was small, histologic results showed that percentage tumor cellularity was inversely related to collagen deposition. Tumors with greater percentage tumor cellularity contained more hyperintense material on T2-weighted and STIR images than did the tumor that was only 5% cellular with abundant collagen. This result agrees with those of other investigators [5, 6] who reported a similar correlation between T2-weighted signal intensity and tumor cellularity and a natural evolution of desmoid tumor toward shrinkage with a decrease in T2-weighted signal intensity. However, the small size of biopsy specimens makes correlation with MRI findings prone to error because small sample volumes may not reflect the predominant tumor composition. This problem may explain our finding of moderate and intense contrast enhancement among tumors that contained mixed amounts of cellularity in abundant collagen within biopsy samples.

We found that the evolution of imaging features of tumors on which follow-up was conducted until the end of adjuvant therapy was quite complex. A surprising finding was that several tumors that grew became less intense on T2-weighted and STIR images and enhanced less than tumors that became smaller. This finding runs counter to the prediction that tumors containing abundant collagen would be less metabolically active than those with less collagen and higher percentage cellularity.

Perhaps increased collagen deposition is partially responsible for tumor enlargement in addition to growth that results from the proliferative activity of fibroblasts. We need to confirm our findings in a larger cohort and to more accurately assess tumor metabolic activity using other imaging techniques.

The main limitations of our study were the retrospective nature and relatively small cohort size. Patients in our study were treated with a variety of chemotherapeutic agents, radiation therapy, and surgery, and imaging time points were not prospectively determined or protocol driven. This limitation made it difficult to assess the evolution of MRI features in response to the therapies and precluded meaningful statistical analysis of the correlation between them. Another study limitation was the lack of uniformity of MRI acquisition techniques because some studies were performed at institutions other than ours. This problem may have limited accurate comparison of MRI features such as tumor signal intensity and contrast enhancement both between patients and between imaging examinations for an individual patient.

Despite the limitations, we found that in children the probability of the presence of residual or recurrent desmoid tumor after surgical resection does not appear to be related to sex or age at diagnosis. MRI assessment of baseline tumor involvement of the neurovascular bundle and bone is probably useful for prediction of the presence of residual or recurrent disease after surgery. Baseline MRI assessment of tumor volume; distinctness of margins; and T1-weighted, T2-weighted, and STIR signal intensity and contrast enhancement patterns does not appear useful for prediction of the presence of residual or recurrent tumor. Of the MRI features we investigated in tumors on which follow-up was conducted until the end of chemotherapy and radiation therapy, size alone seems to be the best indicator of the activity of desmoid tumors. Even so, MRI is invaluable in detection of residual and recurrent disease and in monitoring for tumor progression. Large prospective clinical trials with standardized imaging time points are needed for better assessment of the evolution of MRI features of desmoid tumors in response to therapy. In addition, more precise pathologic–radiologic correlation, including pathologic inspection of the entire resected tumor specimen, when achievable, is necessary for full understanding of the significance of MRI signal characteristics relative to the histologic features of desmoid tumor. Because the biologic activity of this tumor cannot be predicted on the basis of demographic, his-

tologic, or conventional MRI features, functional imaging techniques, such as dynamic contrast-enhanced MRI and contrast-enhanced sonography for quantifying tumor perfusion and PET for assessing metabolic activity, may offer valuable insight into the behavior of desmoid tumor. Such information should allow oncologists to better tailor the clinical management of this difficult disease.

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