MR IMAGING OF AN EXTRAABDOMINAL DESMOID TUMOR OF THE SUPRACLAVICULAR AREA

Chate SIVASOMBOON, MD.

ABSTRACT

Extraabdominal desmoids are benign but locally aggressive tumors, some of which have the appearance on MRI as malignant neoplasm. A case of a pathologically-proven extraabdominal desmoid of the left shoulder involving left levator scapulae muscle is presented with discussion of MRI features of the lesion that may help in preoperative radiological diagnosis, along with a review of literature.

INTRODUCTION

Extraabdominal desmoids are rare benign but locally invasive fibrous tumors. One of their common locations is the supraclavicalar region. MR imaging is useful for their evaluation. Although MRI appearances vary, some MRI features may help to differentiate them from other neoplasms. The extent of local invasion is best shown by MRI, even though it might not be fully depicted. We present the MRI findings of a patient with an extraabdominal desmoid of the levator scapulae muscle which had some MRI features that strongly suggested the diagnosis and turned out to have an unsuspected involvement of the adjacent muscles at surgery.

CASE REPORT

A 39-year-old male patient had had left shoulder pain for a few days, and he had noticed a lump in his left shoulder 4 to 5 months previously. Physical examinaton revealed a mass in the medial portion of his left shoulder, approximately 5cm. in diameter. There were no overlying skin changes and no signs nor symptoms of neurovascular involvement were detected. There was no limitation of the movement of the left shoulder. On MRI, a large mass in the left lower neck and medial portion of the left shoulder, measuring approximately 9.6 cm x 2.5 cm x 6.5 cm, was depicted. It involved the left levator scapulae muscle and was situated just underneath the trapezius muscle, with displacement of the surrouding structures. The tumor seemed to have a fairly well-circumscribed border with no obvious invasion of the surrounding structures. There was no neurovascular encasement. On spin-echo T1wt. images, it had low T1 signal intensity similar to muscle, with a small stellate dark area in the posterior inferior portion of the mass (fig. 1,2). Overall it had bright T2 signal intensity on T2-wt. Images, which were acquired with a fast-spin-echo sequence with fat saturation. A stellate dark area corresponding to the one seen on T1-wt. images was conspicuous on T2-wt. images and additional small stellate and reticular areas of dark T2 signal intensity were evident in the superior portion of the mass (Fig 3,4). After 10 ml. of gadolinium injection, most of the lesion showed a strong enhancement, except the dark unenhanced stellate area, seen on the above T1- and T2-wt. Images (fig. 5, 6). The total extent of the mass was clearly depicted on both T2-wt. and contrast-enhanced T1-wt. images.

Department of Radiology, Faculty of Medicine Chiang Mai University, Chiang Mai 50200 Thailand.

A wide excision was performed with the finding that the tumor also involved the adjacent trapezius and supraspinatous muscles. The histopathological diagnosis was aggressive fibromatosis or extraabdominal desmoid and the margins of the resected specimen were clear of tumor cells. No recurrence was detected after a year of follow-up by physical examinations.

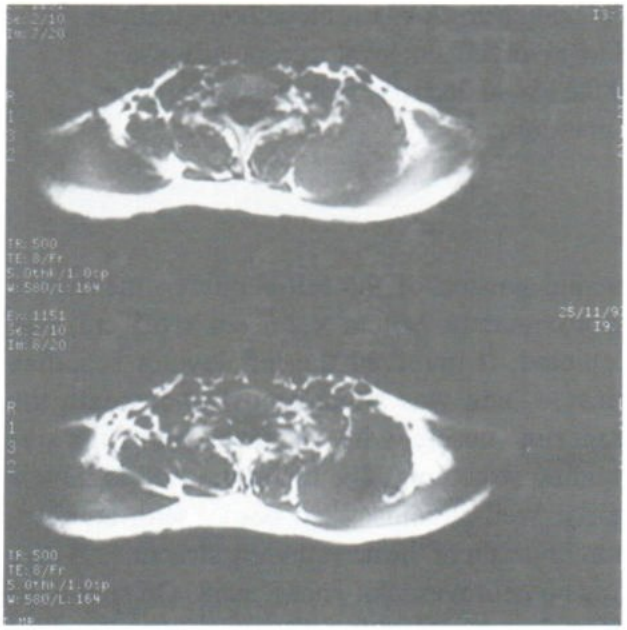

Fig 1. Axial spin-echo T1-wt. images (TR/TE: 500/8) of the supraclavicular regions show a large oval mass in the left supraclavicular region with a signal intensity similar to that of skeletal muscle and containing a stellate T1-hypointese area in the posterior portion (arrowhead).

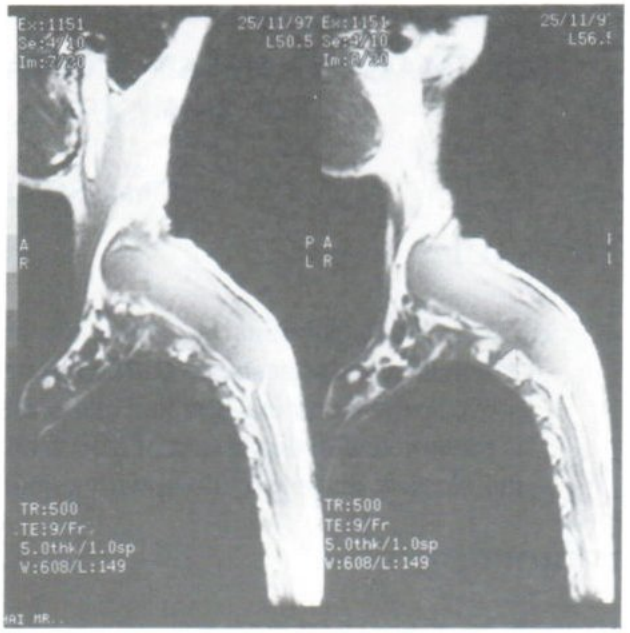

Fig 2. Sagittal spin-echo T1-wt. images (TR/TE: 500/8) of the left supraclavicular region show an elogated mass with well-defined border beneath the trapezius muscle. A dark stellate area is seen in the inferior portion (arrowhead).

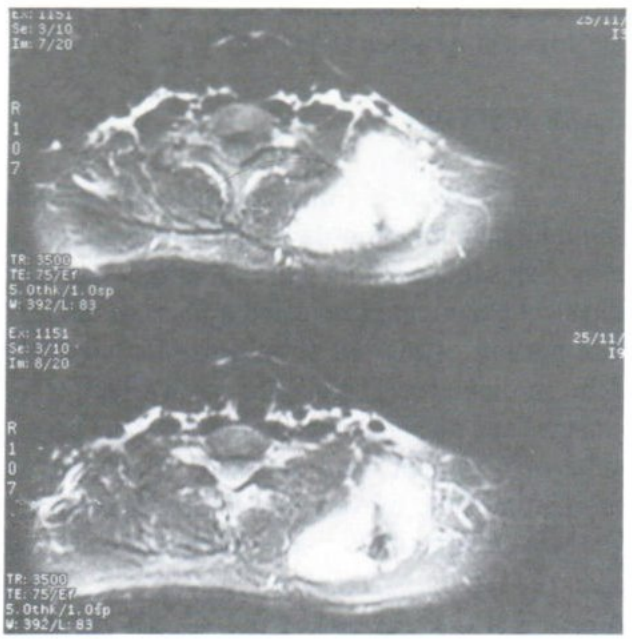

Fig 3. Axial FSE T2-wt. images with fat saturation (TR/TE/ETL: 3500/75/8) in the same locations as the axial T1-wt. images depict a T2-hyperintense mass with a fairly well-defined margin and a prominent stellate dark area in the posterior portion, correponding to the one noted on the axial T1-wt. images.

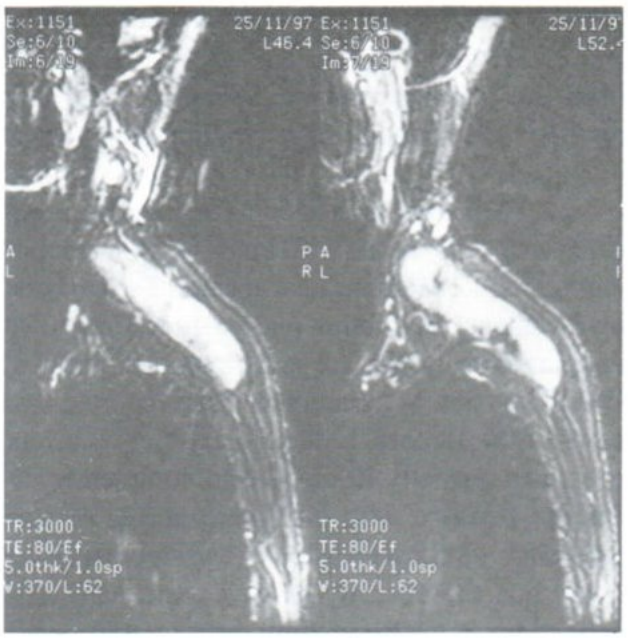

Fig 4. Sagittal FSE T2-wt. images with fat saturation (TR/TE/ETL : 3000/80/8) corresponding to the sagittal T1-wt. images demonstrate a T2-hyperintense mass with a dark stellate area inferiorly and reticular dark areas superiorly. The mass is well-defined.

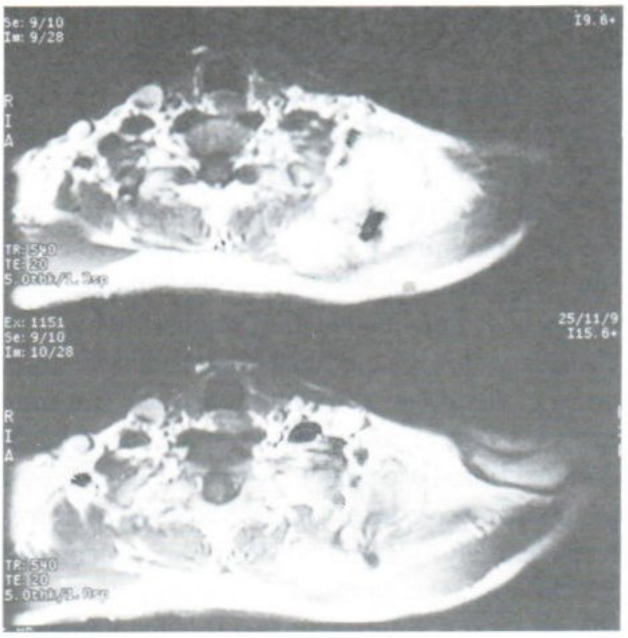

Fig 5. Axial postcontrast T1-wt. images (TR/TE: 540/20) show that the mass in the left supraclavicular region is strongly enhanced with an unenhanced dark area in the posterior portion corresponding to the one seen on the T1- and T2-wt. images.

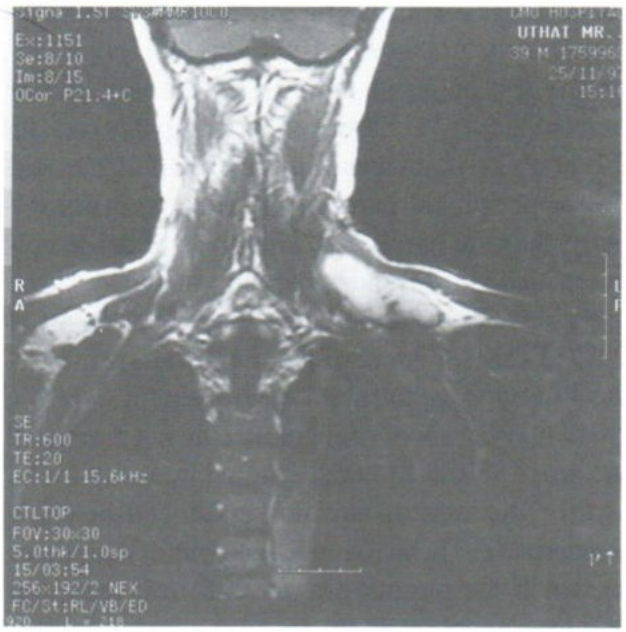

Fig 6. Coronal postcontrast T1-wt. image shows the location of the mass in the supraclavicular region, beneath the trapezius muscle. Note the normal levator scapulae muscle on the contralateral side.

DISCUSSION

Desmoid tumors or aggressive fibromatoses are rare soft tissue tumors which are histologically benign but locally aggressive, arising from connective tissue of muscle, overlying fascia or aponeuroses, and tend to involve the surrounding structures.[1,4,5] They are a subset of a group of lesions called fibromatoses which are classified into (1) superficial (fascial) fibromatoses and (2) deep (musculoaponeurotic) fibromatoses.[2,3,5] Extra-abdominal desmoids or extraabdominal fibromatoses belong to the second group, which also includes two other types of fibromatoses, namely: abdominal desmoids (abdominal fibromatoses) and intraabdominal desmoids (intraabdominal fibromatoses).[2,5,7] Histologically, the tumors show an infiltrative growth pattern and comprise of proliferating fibroblast cells within a matrix of variable amounts of collagen and ground substance, without evidence of cellular anaplasia.[3,4,5]

The incidence of these tumors was previously estimated to be from 3 to 4 per 1,000,000 people per year; they were thought to be much rarer than adominal desmoids.[7] However, data from large pathological series have shown that the overall incidence of extraabdominal desmoids in various anatomical locations is the same as that of abdominal desmoids.[5,6] Seventy percent of the patients are between 25 to 45 years old[5] with female subjects predominating slightly.[6] They are found in every part of the body[6] but usually in the lower limb or pelvic girdle and the shoulder region.[4] The tumors most commonly arise in the intramuscular or intermuscular compartments, but they also occur rarely in the subcutaneous and juxtacortical locations.[4] Multicentricity is rare and usually the tumors are located in the same extremities, oriented along their long axes.[1,3,4] Due to their invasiveness, they tend to recur after resection, recurrence rates being as high as to 65%-70%,[1,4] and these recurrent tumors tend to be more aggressive than the primary ones.[4,5] Some tumors may regress[2,4]; sarcomatous degeneration is very rare.[3]

Their MRI appearances and signal intensities vary, sometimes with invasion of surrounding soft tissue, neurovascular encasement and bone involvement, resembling malignant lesions.[1] The margins of the tumors are sometimes well-defined, sometimes not. Many authors have reported that most of the tumors, both primary and recurrent ones, had partially ill-defined to completely ill-defined margins with high percentages of surrounding soft tissue invasion,[1,2,4] while others have found that the majority of their cases had well-defined tumors,[5,7,8] including the recurrent ones.[5] The tumors usually appear fairly homogenous on T1-wt. images and heterogenous on T2-wt. images.[2,4] On T1-wt. spin echo images, they have a signal intensity ranging from much lower than muscle or equal to fat,[7] but generally the tumors are isointense or slightly hyperintense relative to skeletal muscle.[1,2,4,7] On T2-wt. spin echo images, the predominant signal intensity is typically intermediate between skeletal muscle and subcutaneous fat or isointense with fat, but their signal intensity can be equal to or lower than muscle or higher than fat.[1,2,4,7] Areas of hypointensity on both T1- and T2-wt. images may be present in the central or peripheral parts of the lesions; histologically, they are correlated with areas of dense collagen deposition and hypocellularity.[1,4,5] These areas have been reported to be present in 40 to 90% of cases.[1,5] Most of the tumors show moderate to strong gadolinium enhancement, while a small number show slight or no enhancement, and the areas of low T1 and T2 signal intensity found within the tumors are not enhanced at all.[2,4] Romero et al.[5] have found that in four of their cases in which the tumors did not enhance, they also had low signal intensity on T2-wt. images, probably due to the presence of dense collagen tissues. Although the MRI appearances of the tumors vary,

the presence of these unenhanced low T1 and T2 signal intensity areas within the tumors strongly suggests desmoid tumors or fibromatoses.[1,8] Other malignant tumors such as fibrosarcomas may contain areas of low signal intensity on T2-wt. images, but these areas may not be detected on T1-wt. images and their T2 signal intensity is not as low as it is in desmoid tumors.[1] In case of a recurrent desmoid, it must be differentiated from a post-surgical scar and hematoma.[1,4]

The MRI findings in our case were typical of extraabdominal desmoids or fibromatoses, having well-circumscribed border, T1 isointensity with the muscle, T2 hyperintensity, a strong gadolinium enhancement and a low intralesional T1 and T2 signal area which did not enhance. However, on T2-wt. images,the signal intensity could not be compared with that of fat because of the use of a fast-spin-echo technique with fat saturation. Even though the tumor appeared well-circumscribed, invasion of the adjacent muscles was found at an excision, reflecting the infiltrative growth pattern of the tumor. The patient was also in the common age range and the tumor was located at one of the common sites.[2]

Pathogenesis of the tumors is unclear; there are some factors that may contribute to it, including previous trauma, surgery, radiotherapy and some hormonal factors related to pregnancy.[3,4,5] While intraabdominal desmoids are associated wtih Gardner Syndrome,[5] extraabdominal desmoids have been reported wtih melorheostosis.[9] In juvenile patents with desmoid tumors, an increased frequency of connective tissue anomalies has been noted and these tumors tend to be more aggressive than those occurring in adult patients.[5] The best treatment is en-bloc resection of the tumors, but involvement of important neurovascular structures can prevent complete surgical resection, and recurrence rates of subtotal resection may be as high as 70%.[2,3] Radiation therapy or adjuvant medical therapy may have a role in treating these tumors.[3]

REFERENCES

1. Hartman ET, Berquist TH, Fetsch JF. MR Imaging of Extraabdominal Desmoids: Differentiation from Other Neoplasms. AJR 1992;158:581-585.
2. O'Keefe F, Kim EE, Wallace S. Magnetic Resonance Imaging in Aggressive Fibromatosis. Clinical Radiology 1990;42:170-173.
3. Garant M, Remy H, Just N. Aggressive Fibromatosis of the Neck: MR Findings. AJNR 1997;18:1429-1431.
4. Vandevenne JE, De Schepper AM, De Beuckeleer L, Van Marck E, Aparisi F, Bloem JL. New c oncepts in understanding evolution of desmoid tumors: MR imaging of 30 lesions. Eur. Radiol. 1997; 7:1013-1019.
5. Romero JA, Kim EE, Kim CG, Chung WK, Isiklar I. Different Biologic Features of Desmoid Tumors in Adult and Juvenile Patients: MR Demonstration. J Comput Assist Tomogr 1995;19:782-787.
6. Kransdorf MJ. Benign Soft-Tissue Tumors in a Large Referral Population: Distribution of Specific Diagnoses by Age, Sex, and Location. AJR 1995;164:395-402.
7. Quinn SF, Erickson SJ, Dee PM, Walling A, Hackbarth DA, Knudson GJ, Moseley HS. MR Imgaing in Fibromatosis: Results in 26 Patients with Pathologic Correlation. AJR 1991;156:539-542.
8. Kransdorf MJ, Jelinek JS, Mose RP jr, Utz JA, Judson TM, Neal J, Berrey BI I. Magnetic resonance appearance of fibromatosis: A report of 14 cases and review of the literature. Skeletal Radiol 1990:19: 495-499.
9. Ippolito V, Mirra JM, Motta C, Chiodera P, Bonetti MF. Case report 771: Melorheostosis in association with desmoid tumor. Skeletal Radiol 1993:22:284-288.