DEEP FIBROMATOSIS (DESMOID TUMOR) IN THE TRUNK AND ABDOMEN: CT/MRI AND PATHOLOGICAL FINDINGS

Wichchuda YAMRUBBOON, M.D.[1] Sith PHONGKITKARUN, M.D.[1]
Suphaneewan JAOVISIDHA, M.D.[1] Vorachai SIRIKULCHAYANONTA, M.D.[2]
Jiemjit TAPANEEYAKORN M.D.[1]

ABSTRACT

Objective: To describe imaging findings (Computerized Tomography [CT]/ Magnetic Resonance Imaging [MRI]) correlated with pathological findings, of the deep fibromatosis (desmoid tumor) in the trunk and abdomen.

Material and methods: We retrospectively reviewed such cases in Ramathibodi hospital from January 2001 to December 2005, who had CT/MRI imaging along with pathological slices available.

Results: Seven cases were enrolled in the study ; four of 7 had CT examinations and the other 3 had MR imaging. The locations were extra-abdominal (n=4), intra-abdominal (n=2), and abdominal-wall desmoid tumor (n=1). The margins were well-defined (n=3), partially well-defined (n=3), and ill-defined (n=1). On CT, they were iso- or hypodense. On MRI, although the lesions were iso- to hypointense-T1 and heterogeneously hyperintense-T2 ; there were ill-defined areas of hypointensity in both T1 and T2-weigthed images that was the signal of fibrous tissue. Most lesions showed moderate to strong enhancement after IV contrast injection. Histology showed area of spindle cells in dense collagen fiber, with small & large slit-like vessels. There was no nuclear atypia in majority.

Conclusion: Deep fibromatosis (desmoid tumor) has nonspecific CT findings. In MRI, identifying area of hypointensity on both T1WI and T2WI, with varying degree of enhancement after Gadolinium injection, helps in the diagnosis of the nature of fibrous-origin tumors.

INTRODUCTION

Fibrous-origin tumor is one category of soft tissue tumor that composed entirely of fibrocytes, fibroblasts, myofibroblasts, or mixtures of these three components. Histologically, fibrous tumor may be categorized into benign, intermediate and malignant groups.[1] True incidence is unknown.[2] Majority of cases were found in the extremities (60%), trunk and retroperitoneum (30%).[3] In abdomen, painless mass is the most common sign and symptom and prompts the patients to request imaging studies. CT scan and MR imaging are extremely helpful modalities to visualize tumor details and extension.[4] Unfortunately, definite diagnosis using CT scan alone seems to be difficult.

[1] Department of Radiology and
[2] Pathology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand

The tumors in the intermediate group such as desmoid tumor are quite common. They may behave aggressively mimicking malignancy and cause confusion in the imaging studies. Differentiation between benign and malignant conditions, being very important for management planning, is difficult[3-7] but have to be obtained. Therefore, definite diagnosis is usually based on histological and cytological examinations. In this study, we aimed to review cases in Ramathibodi Hospital and describe the CT/MRI and pathological findings of the deep fibromatosis (desmoid tumor) in the trunk and abdomen.

MATERIALS AND METHODS

The protocol of this study was approved by the Internal Review Board (IRB) and Ethics Committee (EC) of Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand.

Patients

The cases of fibrous-origin tumor were retrieved from surgical pathology reports in the archives of Department of Pathology, Ramathibodi Hospital, from January 2001 to December 2005. According to classification of the fibrous-origin tumor of the Armed Forces Institute of Pathology (AFIP) in 2001.[1] The well known malignancy, malignant fibrous histiocytoma (MFH), was not included in this study because it is a subgroup of fibrohistiocytic tumors.

CT and MRI

The CT scans were performed by using either the 4-slices Multidetector CT (Light Speed plus; GE Medical System, Milwaukee, WI, USA) or the 64-slices Multidetectors CT (Somatom Sensation Cardiac 64; Siemen, Germany).

The upper abdominal CT was performed after administration of the oral contrast medium. The whole abdominal CT was performed after administration of oral and rectal contrast media. Plain axial CT scans were performed with contiguous 3-mm collimation. All enhanced studies were performed after intravenous administration of the Iopromide (Ultravist; Schering Germany) or Iohexol (Omipaque; Amersham, China) with 3-mm slice thickness for upper, lower and whole abdominal CT ; and 2-mm slice thickness for the chest study if there was any.

The MRIs were performed by using 1.5 Tesla machine (CV/I & NV/I; General Electric Medical System, Milwaukee. WI, USA). The MRI sequences for the pelvis consisted of transaxial plane in T1-weightd image (WI), T2WI and T1WI with fat suppression (FS); coronal plane in T1WI and T2WI with FS, and sagittal plane in T2WI with FS. Gadolinium enhanced study was performed by using T1WI with FS in axial and sagittal planes. The MRI sequences for the back included transaxial plane in T1WI, T2WI and T1WI with FS; Coronal plane in T1WI and T2WI with FS, and sagittal plane in T1WI. Gadolinium enhanced study was performed by using T1WI with FS in axial and coronal planes. All sequences were performed with 5.0-mm slice thickness and 1.0 mm gap.

All tumors were reviewed for number, location and size in greatest dimension. The margins were scored as ill-defined, partially well-defined, or well-defined. Presence of hemorrhage, necrosis, calcification, adjacent structural invasion, lymphadenopathy and metastasis were recorded.

For CT scan, we recorded the lesion attenuation as hypodense, isodense or hyperdense. For MR imaging, we recorded the signal intensity relative to the muscle. After intravenous administration of the CT/MRI contrast media, degree of enhancement was recorded as mild, moderate and strong. Pattern of enhancement was recorded as homogeneous and inhomogeneous.

A board certified radiologist and a resident in

Radiology reviewed all images using e-film workstation, version 2.1 (Merge E-film Medical System, USA) on the Window XP operating system (Microsoft Cooperation, USA) with consensus agreement.

Pathology

The reports and microscopic slides of the selected cases were reviewed by a senior pathologist who was an expert in soft tissue tumors in our institute.

RESULTS

There were 115 cases of fibrous-origin tumor in the surgical pathology report of Department of Pathology, consisted of intermediate and malignant groups. Intermediate group (n=80) consisted of desmoid (n=63) and inflammatory pseudotumor (IP) (n=17). The desmoid tumors were found in the head and neck (n=2), extremities (n=32), trunk and abdomen (n=29).

Only seven patients had complete CT (n=4) or MRI (n=3) available. There were 3 males and 4 females, mean age 34.6 years (15-58).

Table 1 summarized data of the patients. Of 7 cases of desmoid tumors, 4 were extra-abdominal (chest wall, back and buttock), 2 were intra-abdominal (mesentery) and 1 was abdominal-wall desmoid. All of them were undergone excisional biopsy for diagnosis and wide excision for treatment. Two cases had recurrence.

The CT or MRI findings

Four patients had CT scans and three had MRI. The mean size was 11.1 cm (range 4.3-15.5). The intra-abdominal group had the largest diameter, averaging 14.0 cm. Three desmoid tumors had well-defined, 3 had partially well-defined, and one had ill-defined margin.

For 4 patients with CT scan, 2 had hypodense attenuation in non-enhanced images, and the rest of tumors had isoattenuation (Fig 1-2)

For 3 patients with MRI, non-enhanced T1WI studies showed variable signal intensity, whereas T2WI showed mixed hypersignal intensity (Fig 3). Two of 3 patients had focal areas of hyposignal in both T1WI and T2WI (Fig 4).

After the administration of the contrast media, all tumors were enhanced in different degrees: minimal (n=1), moderate (n=2) and strong (n=4) (Fig 1-3). The majority of the tumor showed no calcification, hemorrhage, necrosis or adjacent organ invasion. There was no evidence of lymphadenopathy or metastasis.

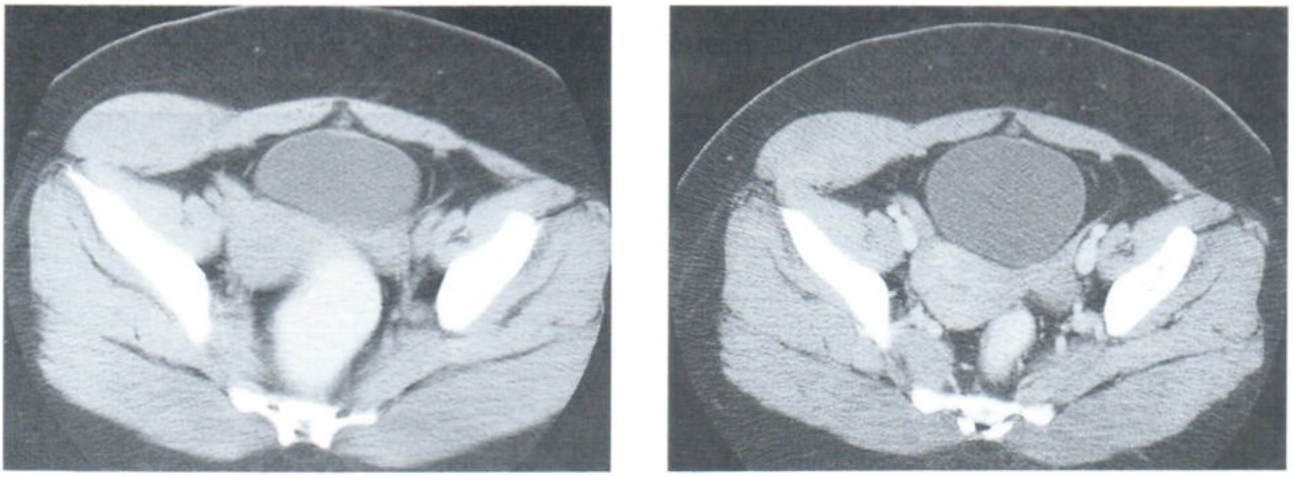
The pathology

The gross pathology revealed mostly ovoid-shape and some irregular-shape masses. All lesions had well-defined or rather well-defined border. The cut surface showed inhomogeneous whitish-tan, whitish-gray or yellow-white tissue with whirling appearance. There were 2 encapsulated masses. Large hemorrhage was seen in 1 case. Histological analysis showed spindle cells in dense collagen fiber. Small and large slit-like vessels were seen. There was no nuclear atypia, except 1 case in the mesentery (case 3) that presented 2-3/50HPF of mitosis. Focal area of fat entrapment was seen in 2 cases. The immunostains were performed in two patients. There was positive reactivity for CD-34 and CD-117 in the lesion at buttock (case 5), and positive reaction for vimentin in the lesion at mesentery (case 3).

Table 1 Summarized CT/MRI and pathological findings of seven cases of desmoid tumors

Case No.	Age (year)	Sex	Location	Mean size (cm)	Number of lesion	CT or MRI findings					Pathological remarks
						Margin	Homogeneity	Attenuation or Signal(SI)	Enhancement	Other findings	
1	28	F	Abdominal wall	10.1	1	Partially well-defined	Inhomogeneous	CT - Isodense	Moderate	-	-
2	44	M	Mesentery	12.6	4	Ill-defined	Inhomogeneous	CT - Isodense	Moderate	Abdominal wall invasion, bowel loop encasement, Hemorrhage, Calcified	-
3	58	M	Mesentery	15.5	1	Well-defined	Inhomogeneous	CT - Hypodense	Strong	Small bowel invasion	Large hemorrhage, Mitosis 2-3/50HPF Immunostain: Positive: Vimentin Negative: CD34, CD68, CD117, S-100, Smooth muscle actin, Desmin
4	32	F	Buttock	4.3	1 with LR*	Well-defined	Homogeneous	MRI - IsoSI T1WI Mixed SI T2WI	Strong	-	Fat entrapment
5	15	M	Buttock	15.0	1	Partially well-defined	Inhomogeneous	MRI - HypoSI T1WI MixedSI T2WI	Strong		Immunostain: Positive: CD34 and CD 117 Negative: Smooth muscle actin -
6	36	F	Chest wall	6.5	1 with LR*	Partially well-defined	Homogeneous	CT - Hypodense	Mild	Pectoralis major and minor muscle invasion Hemorrhage	Fat entrapment
7	29	F	Back	13.4	1	Well-defined	Inhomogeneous	MRI - IsoSI T1WI Mixed SI T2WI	Strong	Hemorrhage	

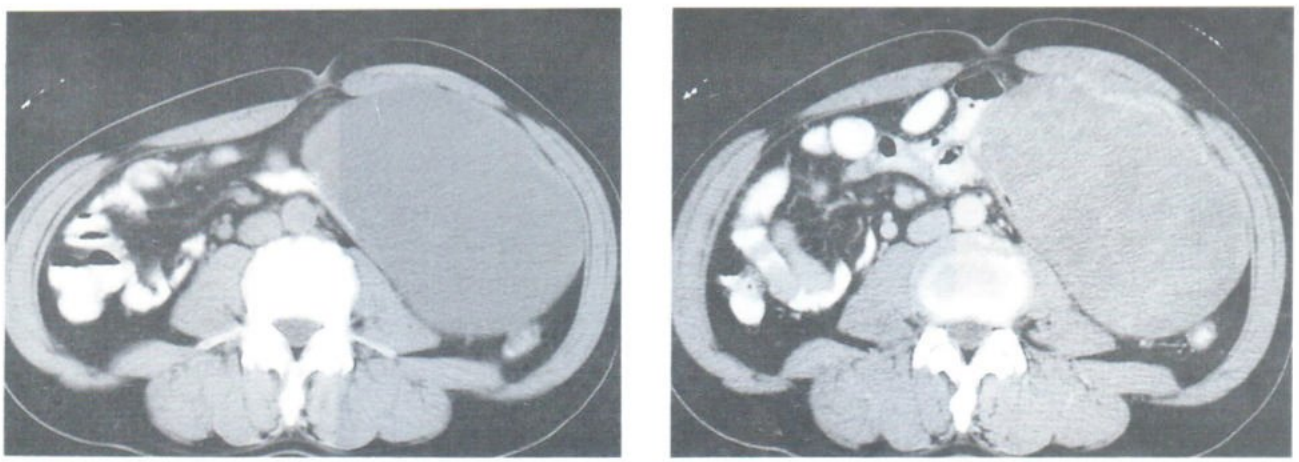
***LR** = Local recurrence



A B

Fig. 1 Case 1; A 28 years old female with abdominal desmoid

Nonenhanced (A) and enhanced (B) CT scans reveal rather ill-defined mixed isodense to hypodense mass with mildly heterogeneous enhancement at the right external oblique muscle.



A B

Fig. 2 Case 3; A 58 years old male with mesenteric desmoid tumor

Nonenhanced (A) and enhanced (B) CT scans reveal a well-defined isodense lesion with heterogeneous enhancement at the mesentery of the left abdomen.

Fig. 3 Case 5; A 15-year-old male with desmoid tumor at the left buttock.

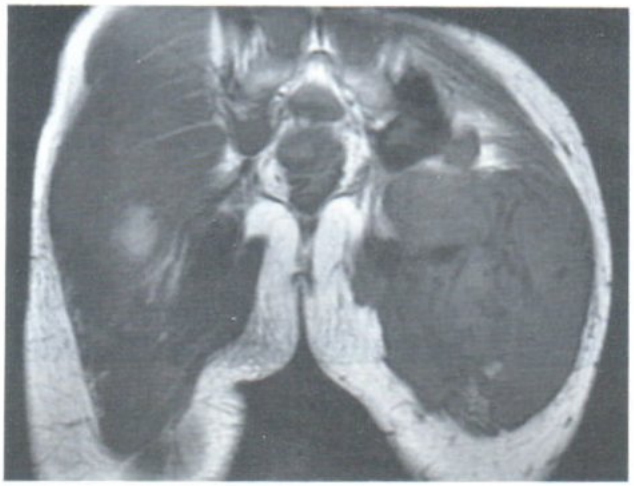


Fig. 3A Coronal T1WI reveals a large, lobulated, ill-defined heterogeneous hyposignal solid mass at the left buttock.

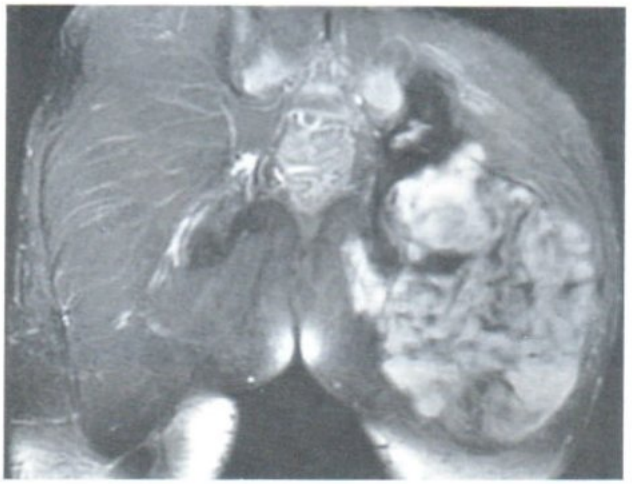


Fig. 3B Coronal T2WI with FS reveals heterogeneous signal of the lesion with multifocal internal low signal areas.

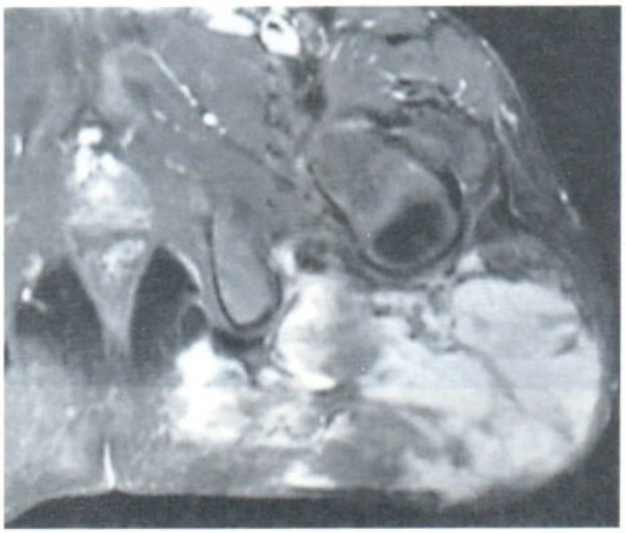
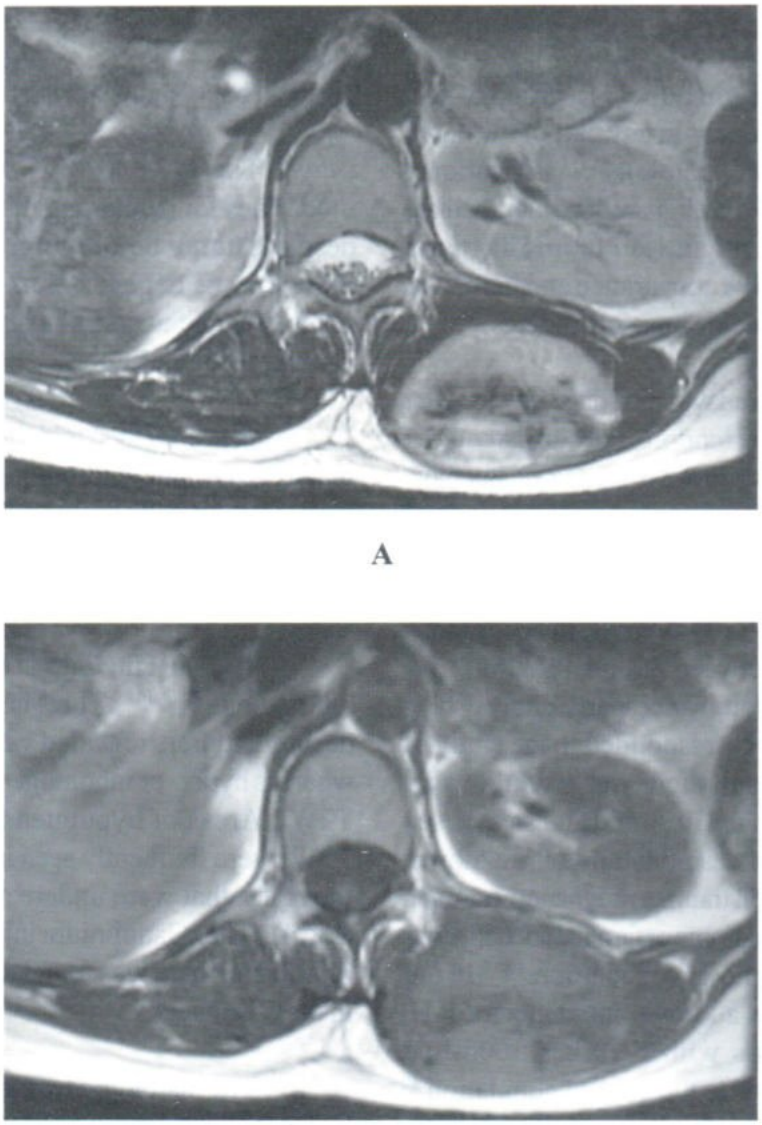


Fig. 3C Axial T1WI with FS after Gadolinium injection reveals no enhancement of multifocal low signal areas in T1WI and T2WI.



A

B

Fig. 4 Case 7; A 29 years old female with desmoid tumor in left side of back

A-B. Axial T1WI (A) and T2WI (B) reveal areas of hyposignal intensity in central portion of the mass in both T1WI and T2WI, representing fibrous area.

DISCUSSION

The fibrous-origin tumor is a group of soft tissue tumor that may occur in any part of the body. They are divided into 3 histology classifications: benign, intermediate, and malignant. The intermediate group is a tumor that might lead to misdiagnosis because of its aggressive appearance and behavior, accompanied by nonspecific imaging findings.

The term desmoid was introduced by Mueller in 1838[8] to describe the tendon-like consistency of the tumor. These tumors do not metastasize, but have a habit of local invasiveness and recurrence after removal are common. The reported incidence is between 3 and 4 cases per million annually.[1-2] The mean age in our study is 34.6 years, in accordance with the literatures (between 25 and 40 years).[9] We also found female predominance, particularly during the reproductive age, corresponded to the study of Kulaylat et al[10] in which there was a relationship between extra-abdominal desmoid and steroid sex hormone. The cause of desmoid tumor is unknown; but surgical or accidental trauma, pregnancy, estrogen hormones, Gardner's syndrome or familial adenomatous polyposis (FAP) are known association.[10-12] In our study, one patient has a known incidence of FAP (case 2). The patients with FAP has 1000-fold increased risk compared to the general population.[10] FAP-associated desmoid tumors occur in intra -abdominal sites in 80-95% of cases, most commonly at mesentery (55-72%) as seen in our study.

The desmoid tumors are traditionally divided, on the basis of anatomical location; into intra -abdominal, extra-abdominal and abdominal-wall desmoid. Intra-abdominal desmoid (intra-abdominal fibromatosis) are those developed in abdominal cavity (e.g. mesentery, retroperitoneum and pelvic cavity). The lesions developed in the abdominal wall, were called abdominal-wall desmoid (e.g. case 1 in Fig. 1). Those lesions originated from deep soft tissue outside the abdomen were classified as extra-abdominal desmoid (extra-abdominal fibromatosis e.g. in the buttock, back, or chest wall).

The CT and MRI were the most useful modalities for evaluating the size and extension of desmoid tumor before surgery. The appearance of these tumors depends on the relative amounts of fibroblast proliferation, fibrosis, collagen contents, and vascularity of the tumor.[13]

In accordance with previous studies, the CT features of desmoid tumor in our study ranged from well-defined to ill-defined. They were isodense or hypodense in noncontrasted CT scan with variable degree of contrast enhancement.[13-15]

The MRI findings in our study correspond to other studies.[8, 14, 16, 17] Three cases of extra-abdominal desmoid tumors revealed variable signal in T1WI, whereas hyper- or inhomogeneous signal are seen in T2WI. Areas of hypointensity in both T1WI and T2WI were frequently seen in the central portion of the mass, that were undetectable from CT images (Fig 4). After Gadolinium injection, variable degree of enhancement was found the same degree as seen in the CT images.

The histologic features were first described by Bennett, as dense fibrous neoplasm arising from the fascial sheaths of striated muscle.[18] On gross histopathologic examination, desmoid tumors were usually circumscribed lesions but they might have irregular or infiltrative border. On cut surface, they were white and coarsely trabeculated, resembling scar tissue. Histologically, desmoid tumors composed of bland spindle or stellate fibroblastic cell embedded in a collagenous stroma, without evidence of muscular or neural differentiation, and with a little or no inflammatory component. They might infiltrate a djacent viscera and tissue at the periphery.[2, 8, 10, 13-15, 17] The cases in our study also presented with similar findings.

Several authors tried to explain MR signals by histological composition. After correlating MRI findings with the histological composition, areas with low signal on both T1WI and T2WI without enhancement represented areas of dense collagen (fibrous component), whereas areas of high signal in T2WI with markedly enhancement represented compact cellular areas. These findings were important for diagnosis of fibromatosis.

The limitations in our study were, first, we excluded more than half of the cases (22 from 29) because of incomplete images or pathological slices. Secondly, due to the inferiority in soft tissue resolution of CT scan, there was no study that correlate exactly the CT attenuation with histologic composition. In our study, the tumor at the abdominal wall (case 1) had a focal area of nonenhancing hypodensity in the central portion, compatible with large area of bleeding, histologically.

In conclusion, deep fibromatosis (desmoid tumor) has nonspecific CT findings. MRI can help both in evaluation and making histological diagnosis due to the signal characteristic of the fibrous tissue. Imaging modalities are found to be useful to evaluate the extension preoperatively, to follow up after treatment, and to detect recurrence if there is any. A study in larger group of patients with complete imaging and histological results would further extend the knowledge about this particular type of tumors.

REFERENCES

1. Kempson R, Fletcher C, Evans H, Hendrickson M, Sibley R. Fibrous and myofibroblastic tumors In: Rosai J, Sobin L, editors. Atlas of Tumor Pathology. Washington, DC. Armed Forces Institute of Pathology 2001 ; 23-104.
2. Enzinger F, Weiss S. General consideration. In: Gay S, editor. Soft tissue tumors. 3 ed. St Louis: Mosby 1995 ; 1-15.
3. Lektrakul N, Chanchairujira K. Imaging of soft tissue tumors. In: Asavamongkolkul A, editor. The Management of Soft Tissue Tumors. Bangkok: P.A.Living 2004 ; 25 - 58.
4. Armstrong S, Watt I. Imaging of soft tissue tumors. Curr Opin Radiology. 1992; 75: 39-44.
5. Sim F, Frassica F, Frassica D. Soft-tissue tumors: diagnosis, evaluation, and management. J Am Acad Orthop Surg. 1994; 2: 202-11.
6. Kransdorf M, Murphey M. Radiologic evaluation of soft tissue masses. AJR. 2000; 175 :575-87.
7. Richard P, JR John E. Radiological evaluation of soft tissue tumor. In: Gay S, editor. Soft tissue tumor. 3 ed. St Louise: Mosby 1995 ; 39-118.
8. Hartman T, Berquist T, Fetsch J. MR imaging of extra-abdominal desmoids: differentiation from other neoplams. AJR. 1992;158: 581-5.
9. Shields C, Winter D, Kirwan W, Redmond H. Desmoid tumors. Eur J Surg Oncol. 2001;27: 701-6.
10. Kulaylat M, Karakousis C, Keaney C, McCorvey D, Bem J, Ambrus S J, et al. Desmoid tumor: a pleomorphic lesion. Eur J Surg Oncol. 1999; 25: 487-97.
11. Clark S, Neale K, Landgrebe J, Phillips R. Desmoid tumors complicating familial adenomatous polyposis. Br J Surg. 1999; 86: 1185-9.
12. Knudsen A, Bulow S. Desmoid tumour in familial adenomatous polyposis. A review of literature. Fam Cancer. 2001; 1:111-9.
13. Casillas J, Sais G, Greve J, Iparraguirre M, Morillo G. Imaging of intra- and extra -abdominal desmoid tumors. Radiographics. 1991; 11: 959-68.
14. Kingston C, Owens C, Jeans A, Malone M. Imaging of desmoid fibromatosis in pedriatric patients. AJR. 2002;178:191-9.

15. Faria S, Iyer R, Rashid A, Ellis L, Whitman G. Desmoid tumor of the small bowel and the mesentery. AJR. 2004;183:118.
16. Vandervenne J, De Schepper A, De Beuckeleer L, Marck E, Aparisi F, Bloem J. New concepts in understanding evolution of desmoid tumor: MR imaging of 30 lesions. Eur Radiol. 1997;7:1031-19.
17. Lee J, Thomas J, Phillips S, Fisher C, Moskovic E. Aggressive fibromatosis: MRI features with pathologic correlation. AJR. 2006;186: 247-54.
18. Rosen R, Kimbell W. Extra-abdominal desmoid tumor. Radiology. 1966;86:534-40.