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# Mesenteric Fibromatosis with Spontaneous Cystic Degeneration: A Case Report with US and CT Findings[1]

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Mesenteric fibromatosis is an uncommon benign neoplasm occurring in the mesentery or retroperitoneum, and presenting as a firm mass with infiltrative margins and homogeneous parenchyma without necrosis or a cystic component (1 - 4). Cystic change may occur, usually after prolonged medical treatment, but is extremely rare (5-7). We describe the US and CT findings in a case of mesenteric fibromatosis with spontaneous extensive cystic degeneration.

**Index words :** Fibromatosis
Desmoid
Mesentery, CT

Mesenteric fibromatosis is an uncommon tumor that affects the mesentery or retroperitoneum. It may arise spontaneously, secondary to previous trauma, surgery, or hormonal stimulation, or in association with a systemic connective tissue anomaly (1 - 3). In fibromatosis, the lesion is usually large and there is extensive infiltration of adjacent structures. Several reports have detailed the computed tomographic (CT) findings (3, 5, 6, 8), but those of spontaneous extensive cystic degeneration have not been described. We report a case of mesenteric fibromatosis with spontaneous extensive cystic degeneration occurring in a young patient who had not previously undergone medical treatment and was not a familial polyposis syndrome sufferer.

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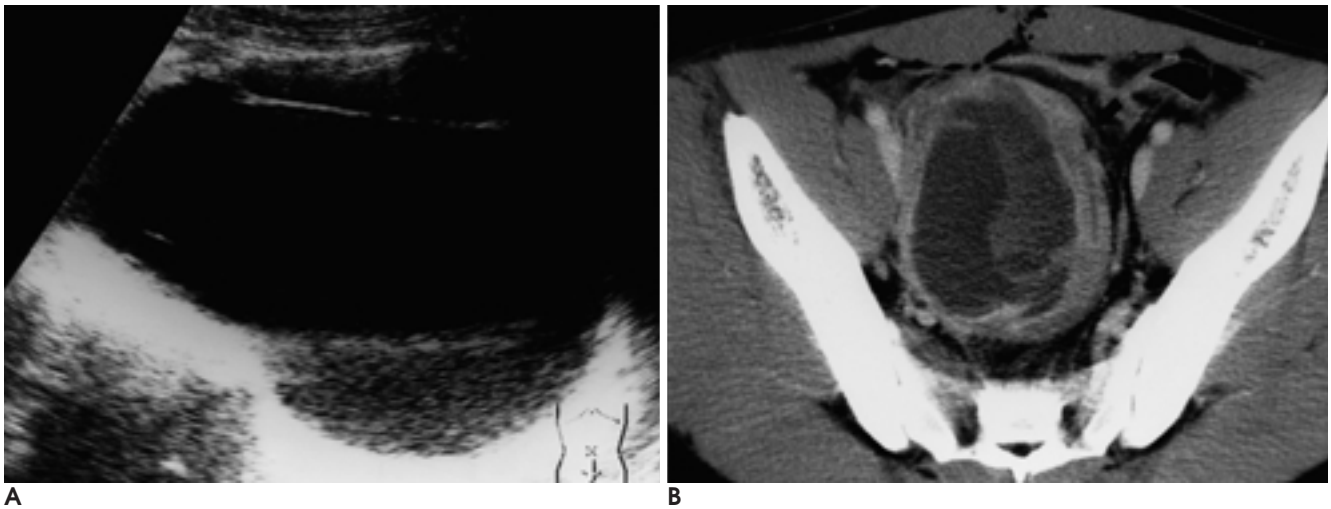
## Case Report

A 20-year-old man whose psat medical and familial history were unremarkable was referred for evaluation of a growing intra-abdominal mass and hydronephrosis of the right kidney. Physical examination disclosed a large, tender, lower abdominal mass.

Ultrasonography revealed that in the lower abdomen, a 10×8×8 ㎝-sized, relatively well-defined, cystic mass with fine septations and soft tissue components was present (Fig. 1A). There was severe hydronephrosis of the right kidney. An unenhanced abdominal CT scan, depicted the mass as focally infiltrating, relatively well marginated and heterogeneous, and various intratumoral attenuations were noted. An enhanced abdominal CT scan showed a large, cystic, solid mass in the pelvic cavity, encroaching on the right mid-to-distal ureter, and at its periphery, strongly enhanced solid components were present (Fig. 1B). Some of the distal small bowel and accompanying mesenteric fat were close to the solid portion of the mass; focal gaseous distention of the bowel had occurred, but neither wall thickening nor en-

hancement were observed. The boundary between the mass and adjacent mesenteric fat was indistinct, and pelvic lymphadenopathy was absent. The mass was thought to be an an unusual type of dermoid tumor or mesenteric cyst.

Surgery revealed that the tumor was firmly attached to the ileum and right distal ureter, and infiltrated the mesenteric fat. The right distal ureter was embedded in the fibrotic wall of the mass, and the proximal ureter was dilatated. The mass, including a segment of the ileum, the right lower ureter and the right colon, was totally excised, and the patient also underwent a ureteroneocystostomy. Gross examination disclosed a 12×8×8 ㎝-sized whitish-gray cystic and solid mass which showed extensive cystic degeneration and had hard elastic soft tissue components. Microscopic examination showed that the lesion was a cystic mass composed of fibroblasts in collagenous stroma of varying density (Fig. 1C). The tumor cells had tapered cytoplasmic processes and uniform, bland nuclei. There was no mitotic activity. Dystrophic calcification and hemorrhage were noted, and the area of cystic degeneration was infiltrated by inflammatory cells. Immunostaining showed the tumor cells to be strongly and diffusely positive for vimentin. The histopathologic and immunohistochemical findings were consistent with those of mesenteric fibromastosis.



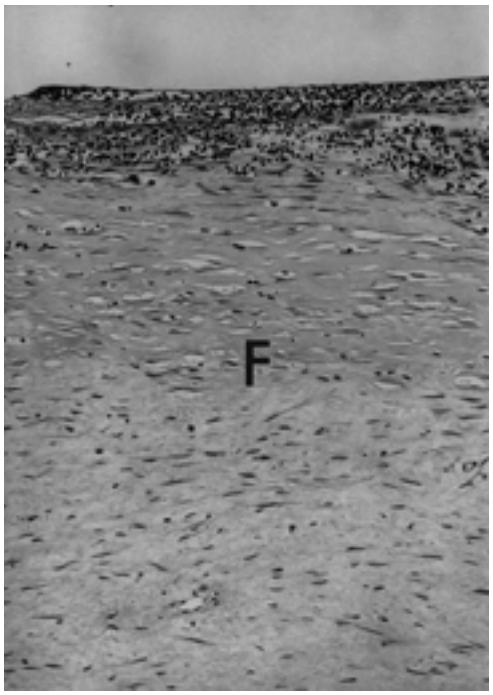




**Fig. 1.** A 20-year-old man with mesenteric fibromatosis, showing spontaneous extensive cystic degeneration in lower abdomen.
**A.** Sagittal US demonstrates pure cystic components with linear septation in upper portion of the mass, and echogenic debris-appearanced soft tissue lesions in deep portion. Posterior sonic enhancement is noted.
**B.** An enhanced CT scan shows central non-enhancing cystic lesion and well enhancing soft tissue components at its periphery. In left side of the mass, the border between the mass and adjacent mesenteric fat is not clearly defined (arrow). At surgery, the mass and mesenteric fat was firmly attached at this level.
**C.** Light microscopic examination demonstrates cystic tumor composed of fibroblasts (F) in the collagenous stroma (hematoxylin-eosin stain, ×100).

## Discussion

The term ' fibromatosis ' covers a broad range of benign fibrous tissue proliferation that has a similar microscopic appearance and is intermediate in its biological behavior between fibrous lesion and fibrosarcoma (1 - 4). Fibromatosis is locally aggressive, showing no features of inflammatory response or malignancy (1 - 4). Histologically, the lesion consists of well-differentiated collagen and fibrous tissue, forming non-encapsulated, poorly circumscribed masses (8). According to its location, fibromatosis is usually divided into two major groups: superficial (fascial) and deep (musculoaponeurotic). Mesenteric fibromatosis (or intra-abdominal desmoid tumor) is a subdivision of deep fibromatosis (2, 3).

Fibromatosis occurs in the mesentery of the small bowel or the retroperitoneum, or at both sites as separate tumors. Less commonly it originates from the ileocolic mesentery, gastrocolic ligament, or omentum. Most mesenteric fibromatoses are large, the majority measuring 10 ㎝ or more in their greatest diameter (3). They are characterized by insidious and invasive growth that may lead to serious complications such as intestinal obstruction, perforation, infarction, or hydronephrosis caused by compression of the small bowel, blood vessels, or ureters (3, 7, 9, 10).

The CT findings of mesenteric fibromatosis have not been extensively reported in the imaging literature (3, 5, 6, 8). Most reported cases have involved lesions with solid soft-tissue elements; in most cases, attenuation was iso- or high (relative to muscle) and margins were either ill- or well-defined. Einstein et al. (5) reported that while the CT appearance of most mesenteric fibromatoses was homogeneous, a variety of heterogeneous patterns was also found, including a low-density center with a high-density rim and a whorled, striped appearance. Necrotic or cystic degeneration was rare (5, 6), usually occurring after prolonged medical therapy. Nakada et al. (7) reported a case involving extensive cystic degeneration after the condition regressed: the patient underwent surgery, after which familial adenomatous polyposis with colon cancer was diagnosed, and was treated with prednisolone for two years. In our case, however, the patient had not been previously treated, and extensive cystic degeneration developed spontaneously.

Mesenteric fibromatosis may occur more frequently in patients with Gardner 's syndrome (3), but the CT features (of margins, attenuation numbers, or response to contrast material), which would permit differentiation between such cases and isolated mesenteric fibromatoses have not been ascertained (3). There are no specific imaging features to distinguish fibromatosis from other solid masses in the abdomen, but in patients with an abdominal mass, a history of abdominal surgery or injury, or Gardner 's syndrome, fibromatosis is a possible diagnosis (6). As in our case, spontaneous cystic degeneration cannot rule out the possibility of mesenteric fibromatosis.

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# 자발적 낭성 변화를 보인 장간막 섬유종증: 초음파 및 CT 소견 1예 보고[1]

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장간막 섬유종증은 장간막이나 후복막에서 생기는 흔하지 않은 양성 종양으로, 대개 낭성 또는 괴사성 변화가 없이 균질한 내부구조를 보이면서 침윤성 병변을 갖는 단단한 종양이다. 낭성 변화는 대개 상당 기간 동안 내과적 치료를 받았던 경우에 일어날 수 있는 것으로 되어있는데, 매우 드문 것으로 알려져 있다. 저자들은 자발적인 낭성 변화를 보였던 장간막 섬유종증 1예를 경험하여 초음파 및 CT 소견을 보고하고자 한다.