

A Solitary Fibrous Tumor of the Retroperitoneum : CT Findings¹

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The solitary fibrous tumor is a rare spindle cell neoplasm of mesodermal origin. It usually arises in the pleura, though has recently been reported in the peritoneum and at nonserosal sites; it is, however, extremely rare in the retroperitoneum. The authors describe one case of a solitary fibrous tumor that occurred as a huge complex mass in the retroperitoneum of an adult. Preenhanced CT scans showed that the mass was sharply delineated from surrounding organs and that tumor density was the same as that of surrounding musculature. Peripheral Calcifications were noted, and postenhanced scans showed intense, gradual enhancement of solid portions of the mass, with nonenhanced cystic portions. Although not specific, solitary fibrous tumors must be included in the differential diagnosis of a large retroperitoneal tumor of complex consistency and with internal calcification.

Index words : Retroperitoneal space, CT
Retroperitoneal space, neoplasms

A solitary fibrous tumor is a rare spindle cell neoplasm of mesodermal origin. It usually arises in the pleura, though has recently been reported in the peritoneum, and at nonserosal sites; it is, however, extremely rare in the retroperitoneum(1. 2). We described the CT findings of a solitary fibrous tumor that occurred there as a huge complex mass; the findings are correlated with those of pathology.

Case Report

A 27-year-old woman presented with a 6-year history of vague, ill-defined abdominal pain and marked abdominal distension. She had experienced neither loss of appetite nor weight loss. Six years previously, ovarian cancer with no possibility of cure had been diagnosed at another hospital, but the patient had not

undergone abdominal surgery. Routine hematological and biochemical tests including liver function tests and urinalysis were normal. A plain radiograph of the abdomen obtained with the patient upright showed a vague, huge soft tissue mass with a few calcifications and peripherally displaced gas-containing bowel loops.

Subsequent CT scans showed an enormous, well-defined mass filling the peritoneal cavity with ascites. It was sharply delineated from surrounding organs other than the left kidney(Fig. 1A), and enhanced CT scans showed that it consisted of intensely enhanced solid portions and nonenhanced round cystic portions(Fig. 1 A, B). Calcifications were seen in the periphery of the tumor, and unenhanced scans, obtained from the other hospital, showed apparent intermediate attenuation. Because of its huge size and inhomogeneous complex consistency, the preoperative diagnosis was primary retroperitoneal sarcoma such as malignant fibrous histiocytoma, leiomyosarcoma, or pleomorphic liposarcoma.

Because diagnosis was uncertain, the patient underwent an exploratory laparotomy, and a huge, well capsulated and lobulated, pinkish white glistening mass with a stalk was found in the area of the

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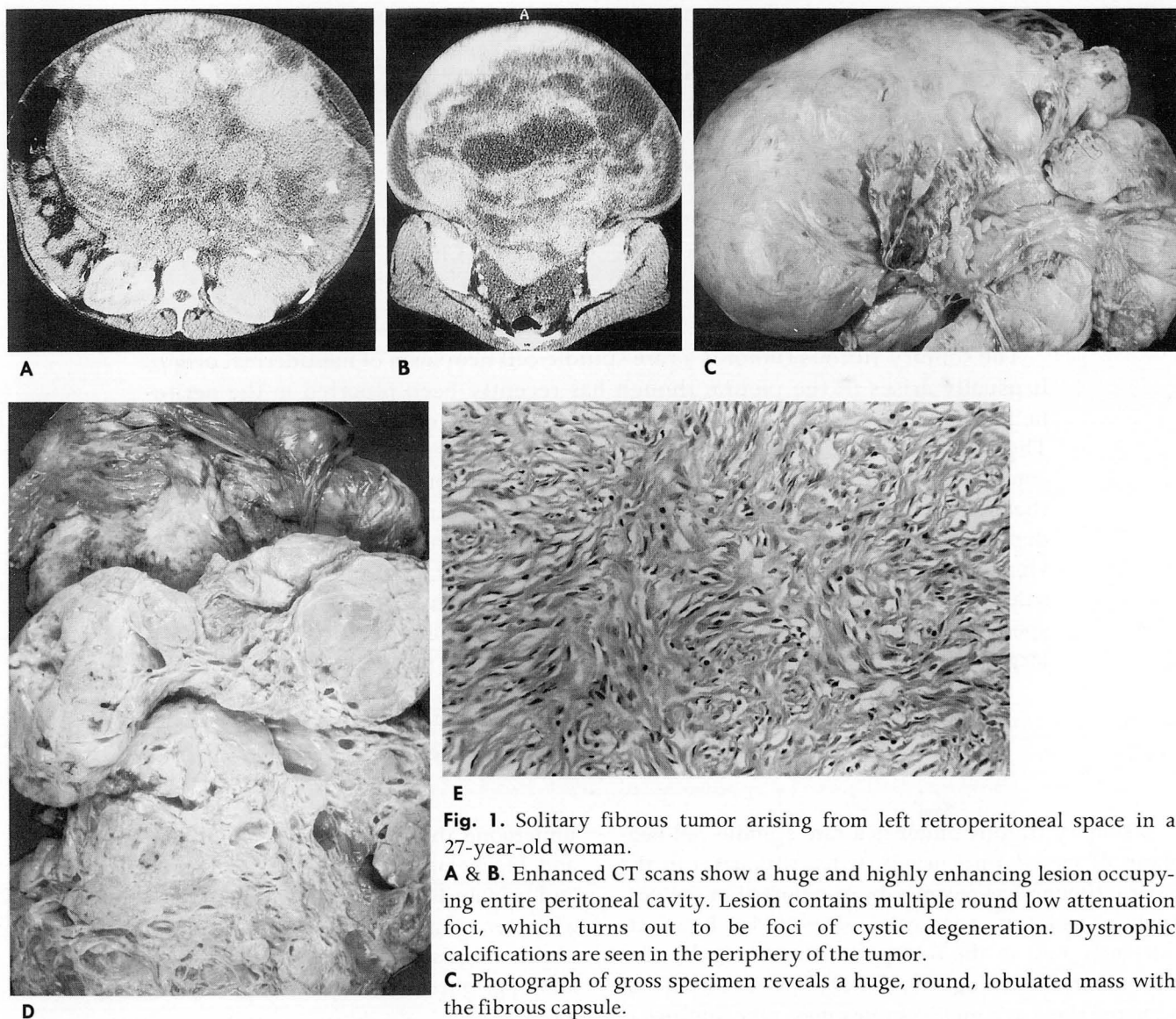


Fig. 1. Solitary fibrous tumor arising from left retroperitoneal space in a 27-year-old woman.

A & B. Enhanced CT scans show a huge and highly enhancing lesion occupying entire peritoneal cavity. Lesion contains multiple round low attenuation foci, which turns out to be foci of cystic degeneration. Dystrophic calcifications are seen in the periphery of the tumor.

C. Photograph of gross specimen reveals a huge, round, lobulated mass with the fibrous capsule.

D. Cut slice of gross specimen reveals a yellowish white fibrotic lesion intermingled with multiple foci of cystic degeneration.

E. Photomicrograph of the specimen shows spindle cells having slender and elongated cytoplasmic process in the collagenous background (Hematoxylin and Eosin. $\times 400$).

retroperitoneum near the left kidney. Evaluation of a surgically excised pathologic specimen revealed a round mass, measuring $50 \times 40 \times 20$ cm and weighing 2400 grams, with a capsule (Fig. 1C). The cut surface of the gross specimen showed a yellowish white fibrotic lesion intermingled with multiple foci of cystic degeneration that contained yellow serous fluid (Fig. 1D). Microscopic examination indicated that the main solid portions were areas of mixed cellular and collagenous tissue (Fig. 1E). Enhanced scanning revealed high attenuation, and on CT scans, areas of cystic degeneration on gross specimen manifested as areas of round, cystic, fluid attenuation. Microscopic examination showed no mitosis or cellular dysplasia. Immunologic staining of tumor cells, however, showed

a positive reaction with CD34 and vimentin, but cytokeratin and S100 protein consistently reacted negatively, thus confirming that the mass was a solitary fibrous tumor simulating the counterpart of pleural origins.

Discussion

Solitary fibrous tumors, also called benign fibrous mesotheliomas, are rare tumors of mesodermal origin. They primarily affect the pleura and occasionally occur in the peritoneum and at nonserosal sites (1–3). Reports of their CT findings have been very limited, even in pleural lesions (4–6). CT findings of solitary fibrous tumor of the pleura include well-delineated,

often lobulated, noncalcified, enhancing soft-tissue masses in close relation to the pleural surface or fissure, associated crural thickening, and an absence of chest wall invasion.

Yu et al.(7) reported one case of CT findings of a solitary fibrous tumor in the retroperitoneum. The presented CT findings of that case were a large, well-delineated, solid mass, with strong contrast enhancement, and a lack of massive necrosis or calcification. Evaluation of the specimen revealed a huge, round, solid mass of homogeneous consistency and without a definite capsule. In our case, however, the radiological findings of the tumor were different from those of the previously reported case. The features which distinguished it from our case were multiple areas of cystic degeneration, calcifications in the peripheral portion of the mass, a definite capsule, and a pedicle.

Briselli et al.(2) reported that small solitary fibrous tumors of the pleura were nodular and homogeneous in appearance, but in large lesions, foci of degeneration and cystic change were common. In nine patients, Lee et al.(4) correlated CT findings of benign fibrous mesothelioma of the pleura with pathologic findings. The significant enhancement always seen on enhanced CT scans has been explained by the vascularity of the tumor, which frequently shows myxoid and sometimes contains cystic degeneration, and may contain a hemorrhage, as seen in the low density or non-enhanced portion. These features were very similar to those of our case. In addition, Hutchinson and Friedenber(8) observed dystrophic calcification in two of 17 cases of benign fibrous mesothelioma of the pleura, and we thus believe that calcification of the tumors may not be rare. We believe that all the features of solitary fibrous tumors of the retroperitoneum are the same as those of tumors of the pleura, and that

differences in gross and microscopic features of the two tumors found in the retroperitoneum might be growth-related.

The CT findings in this case are not pathognomonic but help narrow the differential diagnosis, which for a solitary fibrous tumor of the retroperitoneum includes malignant fibrous histiocytoma, pleomorphic liposarcoma and leiomyosarcoma. Findings of a well-defined margin without local invasion or distant metastases in spite of its huge size, and the presence of dystrophic calcification and strong enhancement of the solid portion, with nonenhanced cystic portions, as seen on CT, may help differentiate this tumor from other primary retroperitoneal neoplasms.

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후복막강에서 기원한 고립 섬유성 종양의 전산화 단층촬영 소견: 1예 보고¹

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고립 섬유성 종양은 주로 늑막에서 발생하는 중배엽기원의 종양으로 간혹 복막이나 장막에 싸여 있지 않은 부위에서도 간혹 보고가 있었으나, 후복막에서 발생한 고립 섬유성 종양은 매우 드물다. 저자들은 후복막강내에서 발생한 고립 섬유성 종양 1예를 경험하였기에 CT소견을 병리소견과 비교분석하여 보고한다. 조영전 CT상 종괴는 비교적 명확한 경계를 보였고 주변 근육과 비슷한 정도의 불균질한 밀도를 가지고 있었으며 일부에서 작은 석회화가 관찰되었다. 조영후 CT상 종괴는 복막강내를 거의 완전히 차지하는 대형 종괴였으며, 강한 조영증강을 보이는 고형 부분과 주변의 복수와 비슷한 밀도를 가진 조영증강이 되지 않는 낭성부분이 섞여 있는 매우 불균질한 모양을 보였다. 비특이적이긴하나 후복막강에 석회화를 동반한 낭성병소와 고형병소가 섞여있는 대형종괴가 있을 때 이 질환을 감별진단의 하나에 포함시켜야 할 것으로 생각된다.