Korean J Gastroenterol Vol. 57, No. 1, 47-50
DOI: 10.4166/kjg.2011.57.1.47

CASE REPORT

KJG

위장의 점막하종양과 유사한 후복막 악성 고립성 섬유종

배정민, 김세원, 김상운, 송선교
영남대학교 의과대학 외과학교실

Malignant Solitary Fibrous Tumor of Retroperitoneum Mimicking Gastric Submucosal Tumor

Jung Min Bae, Se Won Kim, Sang Woon Kim and Sun Kyo Song
Department of Surgery, Yeungnam University College of Medicine, Daegu, Korea

Solitary fibrous tumors (SFTs) are an uncommon neoplasm characterized by the proliferation of spindle cells. The diagnostic criteria of malignant solitary fibrous tumors (MSFTs) include high cellularity, high mitotic activity (4＞10 HPF), pleomorphism, hemorrhage and necrosis. This tumor frequently involves the pleura and MSFTs of retroperitoneum mimicking gastric submucosal tumor are very rare. We report a rare case of MSFT that presented as a gastric submucosal tumor. A gastroscopic examination showed a large bulging mucosa in the gastric body. Abdominal computed tomography revealed a well-defined heterogeneous enhancing mass between the left hepatic lobe and gastric body. Surgical resection was performed and histologic features were consistent with a MSFT. **(Korean J Gastroenterol 2011;57:47-50)**

Key Words: Malignant solitary fibrous tumor; Retroperitoneum; Gastric submucosal tumor

INTRODUCTION

Solitary fibrous tumors (SFTs) are rare neoplasms characterized by proliferation of spindle cells that usually affect the pleura, with malignant form being rarer than the benign form. SFTs are diagnosed pathologically by the presence of spindle cells exhibiting diffuse CD34 and bcl-2 positivity on immunohistochemical analysis. The diagnostic criteria of malignant solitary fibrous tumors (MSFTs) include high cellularity, high mitotic activity (>4/10HPF), pleomorphism, hemorrhage, and necrosis.[1]

Fibrosarcomas, malignant fibrous histiocytomas, GISTs, hemangiopericytomas, synovial sarcomas, and malignant mesenchymomas should be included in the main differential diagnosis of solitary fibrous tumors.[2] SFTs has also been reported in other anatomic sites, including the peritoneum, pancreas, paranasal sinuses, nose, and pericardium. Due to such a variety of anatomic sites, clinical manifestations can vary according to the location and size of the mass. Case with malignant solitary fibrous tumor of the retroperitoneum mimicking a gastric submucosal tumor is extremely rare and has not been cited in the current literature. We report a rare

Received January 26, 2010. **Revised** May 7, 2010. **Accepted** May 7, 2010.

교신저자: 김상운, 705-030, 대구시 남구 대명동 317-1, 영남대학교 의과대학 외과학교실
Correspondence to: Sang Woon Kim, Department of Surgery, Yeungnam University College of Medicine, 317-1, Daemyung-dong, Namgu, Daegu 705-030, Korea. Tel: +82-53-620-3580, Fax: +82-53-624-1213, E-mail: swkim@med.yu.ac.kr

Financial support: None. Conflicts of interest: None.

case of MSFT that presented as a gastric submucosal tumor.

CASE REPORT

A 59-year-old man presented with a gastric submucosal tumor detected incidentally during a routine health screening. He had no known family history of gastrointestinal disorders or cancers. On admission, he had neither abdominal discomfort nor any symptom. Physical examination on admission revealed no abnormalities. The initial biochemical and hematologic investigation, including complete blood count, electrolytes and liver function tests were all within normal range. A chest radiograph finding showed no abnormalities.

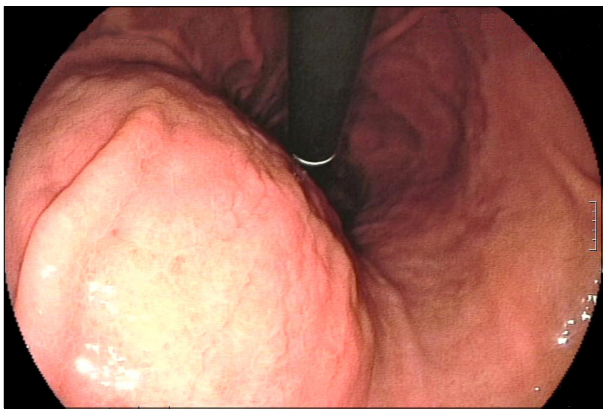

Fig. 1. Gastroscopic finding showed huge bulging mucosa in the lesser curvature side of the gastric body.

A gastroscopic examination showed a large bulging mucosa in the lesser curvature side of the gastric body (Fig. 1). Non-contrast and contrast abdominal computed tomography (CT) revealed a well-defined heterogeneous enhancing mass, about 10 cm in size between the left hepatic lobe and the lesser curvature side of the gastric body (Fig. 2), suggesting submucosal tumor, such as a GIST, schwannoma, or leiomyoma. No evidence of distant metastasis or significant lymph node enlargement was noted. Surgical resection was performed, and the mass was located between the left hepatic lobe and the lesser curvature side of the gastric body. The tumor was adherent to the gallbladder and retroperitoneum, but gastric wall was intact with no tumor invasion or wall deformities and there was no evidence of gastric submucosal lesion. Considering those findings, origin of the present tumor was assumed as the retroperitoneum. The tumor was excised successfully with a negative resection margin.

The tumor was 22.0×12.0×5.0 cm in size (Fig. 3). The cut surface showed a gray-white to yellow-white solid mass with a focal area of cystic change and necrosis. The spindle-shaped cells were positive for CD34, Bcl-2, and CD99 (Fig. 4). Immunoreactivity for desmin and S-100 protein was negative. The mitotic activity was 10 per 10 HPF. There was no vascular invasion. Histologic features were consistent with a MSFT.

The post-operative course was uneventful, and he was doing well following surgery. No evidence of re-

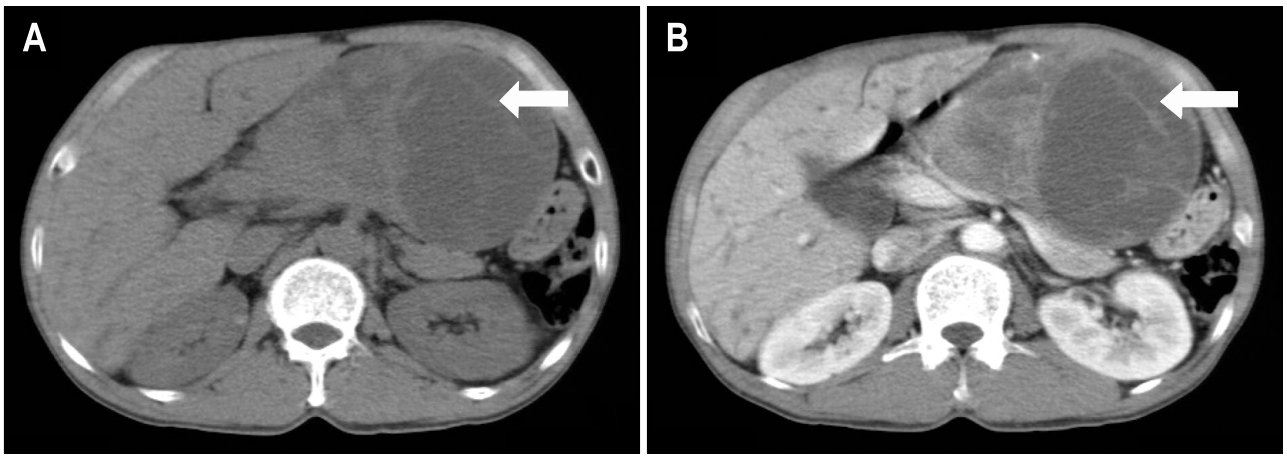

Fig. 2. Abdominal CT findings. (A) Non-contrast abdominal CT showed a heterogenous mass like lesion (white arrow). (B) Contrast abdominal CT showed a well-defined heterogeneous enhancing mass, about 10 cm in size between the left hepatic lobe and the lesser curvature side of the gastric body (white arrow).

currence or metastasis was observed at the 36 month follow-up.

DISCUSSION

SFTs are rare neoplasms that usually affect the pleura.[1] On occasion, extrapleural SFTs have been reported in the literature. Extrapleural lesions include the peritoneum, pericardium, anus, and paranasal sinuses.[3] The benign form is three-to-four times more common than the malignant form.[4] A MSFT of the retroperitoneum mimicking gastric submucosal tumor has not been reported in the literature.

Fibrosarcomas, malignant fibrous histiocytomas, GISTs, hemangiopericytomas, synovial sarcomas, and malignant mesenchymomas should be included in the main differential diagnosis of solitary fibrous tumors.[2] The pathogenesis of SFTs remains unknown.

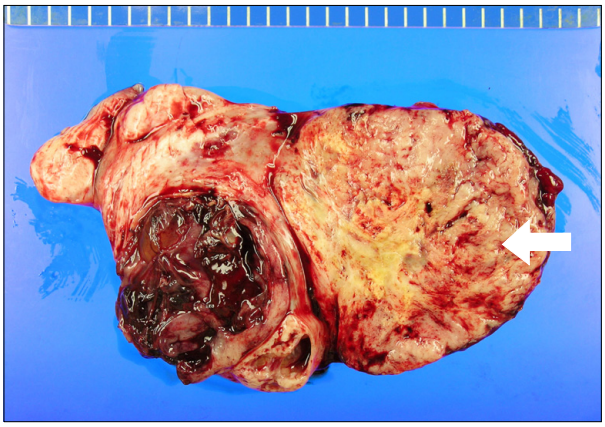

Fig. 3. Gross appearance of cut surface showed yellowish white solid mass with focal area of cystic change and necrosis (white arrow). The tumor was 22.0×12.0×5.0 cm in size.

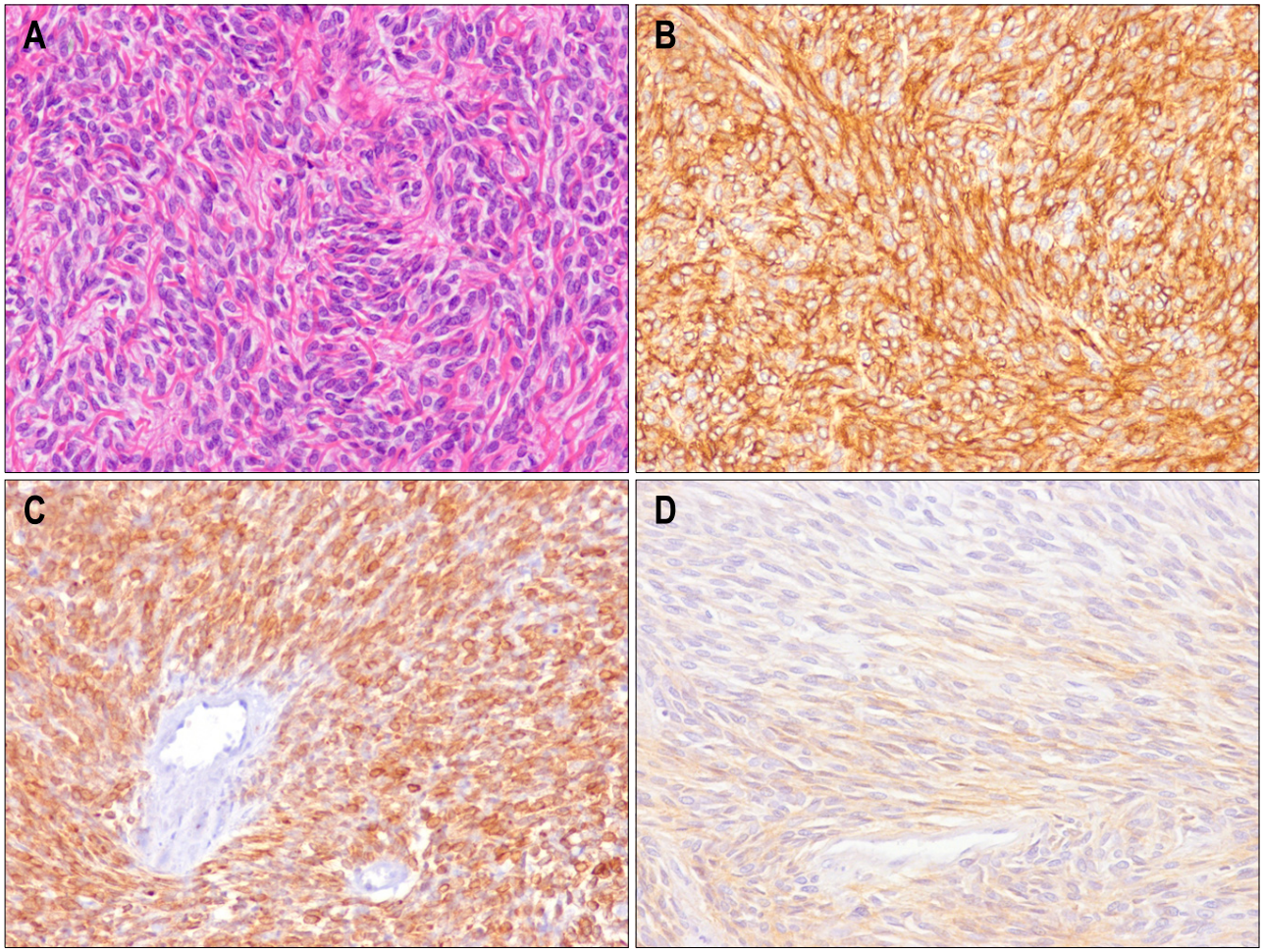

Fig. 4. Histopathologic findings. (A) This histopathologic features were consistent with spindle shaped cells (H&E, ×200), (B) The tumor cells showed strong positive staining for CD34 (×200), (C) The tumor cells showed strong positive staining for Bcl-2 (×200), (D) The tumor cells showed focally weak positive staining for CD99 (×200).

The clinical presentation of SFTs varies greatly, depending on the location and size of the mass. A correct pre-operative diagnosis of SFTs is difficult because clinical manifestations, physical examination findings, and imaging results are not distinctive. On computed tomography findings, SFTs appear as well-defined, solid tumors containing cystic portions.[5] In the present case, he had no symptom related to tumor. But, the mass was founded incidentally on gastroscopy during a routine health examination. An abdominal CT revealed a well-defined heterogeneous enhancing mass.

Only histopathologic evidence can allow a correct diagnosis of SFTs. An intact layer of mesothelium overlying the tumor characterizes SFTs histopathologically. The tumor is composed of spindle-shaped cells resembling fibroblasts with varying amounts of hyalinized collagen. SFTs are diagnosed pathologically by the presence of spindle cells exhibiting diffuse CD34 and bcl-2 positivity, and S100, actin, and keratin negativity, on immunohistochemical analysis.[6] The function of CD34 is controversial; however, many authors believe that SFTs arise from ubiquitous CD34-positive interstitial cells.[7] Moreover, the diagnostic criteria of MSFTs include high cellularity, high mitotic activity (>4/10/HPF), pleomorphism, hemorrhage, and necrosis.[1]

The most effective therapeutic modality in SFTs is surgical resection.[8] Recurrence of a SFT as long as 7 years after surgical resection has been documented.[3] Follow-up radiologic examination is necessary at least once a year. For recurrence of peritoneal SFTs, tumor re-excision securing negative surgical margins is recommended.[9] Distant metastases, such as lung, liver, and bone, have been reported in peritoneal SFTs.[8] The value of adjuvant chemotherapy is controversial. The most important prognostic factor in abdominal SFTs is unknown. In case with pleural SFTs, the respectability is the most important prognostic factor, but the histologic features cannot exert influence on disease prognosis.[1] Therefore, we have proposed surgical excision whenever possible.

In conclusion, we present a rare case of MSFT in the retroperitoneum mimicking gastric submucosal tumor and diagnostic modalities for tissue sampling and immunohistochemical study are mandatory in confirming an accurate diagnosis.

REFERENCES

1. England DM, Hochholzer L, McCarthy MJ. Localized benign and malignant fibrous tumors of the pleura. A clinicopathologic review of 223 cases. Am J Surg Pathol 1989;13:640-658.
2. Young RH, Clement PB, McCaughey WT. Solitary fibrous tumors ('fibrous mesotheliomas') of the peritoneum. A report of three cases and a review of the literature. Arch Pathol Lab Med 1990;114:493-495.
3. Hasegawa T, Matsuno Y, Shimoda T, Hasegawa F, Sano T, Hirohashi S. Extrathoracic solitary fibrous tumors: their histological variability and potentially aggressive behavior. Hum Pathol 1999;30:1464-1473.
4. Tanaka M, Sawai H, Okada Y, et al. Malignant solitary fibrous tumor originating from the peritoneum and review of the literature. Med Sci Monit 2006;12:CS95-98.
5. Cardinale L, Allasia M, Ardissone F, et al. CT features of solitary fibrous tumour of the pleura: experience in 26 patients. Radiol Med 2006;111:640-650.
6. Hanau CA, Miettinen M. Solitary fibrous tumor: histological and immunohistochemical spectrum of benign and malignant variants presenting at different sites. Hum Pathol 1995;26:440-449.
7. Chan JK. Solitary fibrous tumour--everywhere, and a diagnosis in vogue. Histopathology 1997;31:568-576.
8. Kishi K, Homma S, Tanimura S, Matsushita H, Nakata K. Hypoglycemia induced by secretion of high molecular weight insulin-like growth factor-II from a malignant solitary fibrous tumor of the pleura. Intern Med 2001;40:341-344.
9. Vallat-Decouvelaere AV, Dry SM, Fletcher CD. Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors. Am J Surg Pathol 1998;22:1501-1511.