

Case Report

Giant solitary fibrous tumor of the diaphragm: a case report and review of literature

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Abstract: A young gentleman presented with difficulty in breathing. Computed tomography (CT) scan showed a huge mass located between the heart and stomach, which might have rooted in the diaphragm. Magnetic resonance imaging (MRI) with enhanced three dimensional construction showed a lobulated, heterogeneous soft tissue mass with short T1 weighted imaging signal and flake long T2-weighted imaging (T2WI). Tumor-enhanced scanning demonstrated heterogeneous contrast enhancement. The preliminary diagnosis was intra-abdominal huge mass and considering sarcoma. Resection was conducted where the base of the tumor was located in the diaphragm oppressing the left liver lobe and heart. The base of the tumor, together with partial surrounding of the diaphragm, pericardium base, and the left lateral hepatic segment, was resected. The defect in the diaphragm and pericardium was repaired by patching, and thoracic close drainage and abdominal drainage were placed following the surgical operation. The pathological report showed giant solitary fibrous tumor (SFT). This case report may provide a reference resource for the diagnosis and treatment of SFT located in the diaphragm.

Keywords: Solitary fibrous tumor, SFT, diaphragm

Introduction

Solitary fibrous tumor (SFT) is a rare spindle cell tumor, which is rarely presented in the diaphragm [1]. Preoperative diagnosis of diaphragmatic SFT is challenging because its morphological features are not well detected by radiological examinations.

This case report aims to provide information on clinical experience, imaging outcomes and pathological features, and treatment of SFT, with the objective of aiding the correct diagnosis and radical resection of the tumor. Written informed consent was obtained from the patient.

Case report

The patient was a 21-year-old male, who was hospitalized with an abdominal mass and a history of difficulty in breathing for one week. He was admitted to a hospital to take a computed tomography (CT) scan, which showed a huge

mass located between the heart and stomach, and which might have located to the diaphragm. Next, he was admitted to our hospital for further treatment. Physical examination showed that all vital signs were stable. A hard mass, with dimensions of 20 × 20 cm was palpable and was located in the upper left abdomen with no clear border separating the tumor from the surrounding tissue (**Figure 1C**). Radiological examination consisted of magnetic resonance imaging (MRI) and showed a lobulated, heterogeneous soft tissue with short T1-weighted imaging and a flake long T2-weighted imaging (T2WI) signal. Tumor-enhanced scanning demonstrated heterogeneous contrast enhancement.

The left lobe of the liver, stomach, pancreas, heart and diaphragm were clearly and obviously compressed (**Figure 1A** and **1B**). The mass adhered to the pericardium, and not to the cardiac muscle (as shown by replayed MRI video). The preliminary diagnosis was that of a huge intra-abdominal neoplasm that was considered

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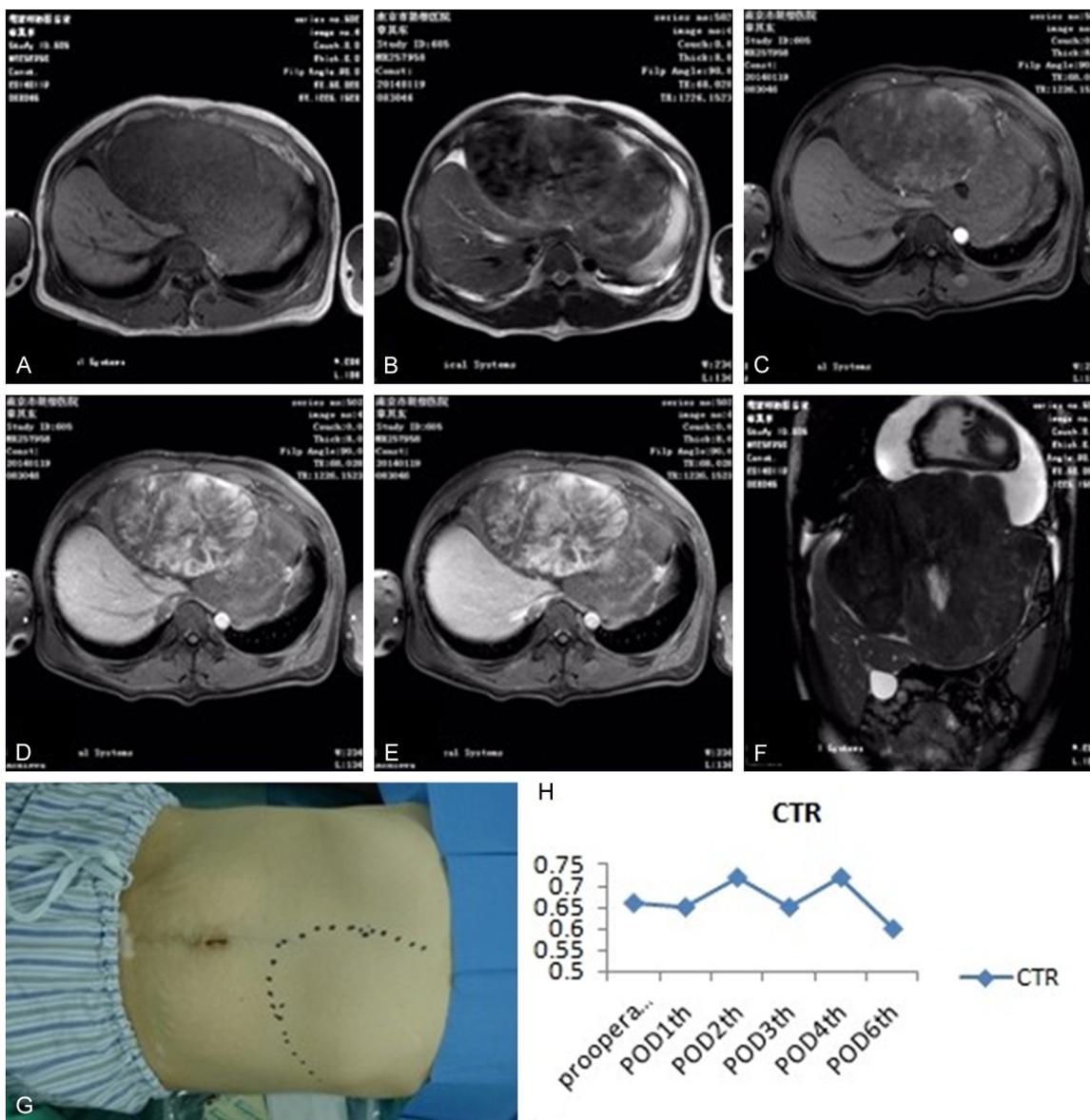


Figure 1. The tumor had dimensions of 22.3 × 19.4 × 12.4 cm and had an uneven density, pressing the surrounding organs such as the liver, heart and others. (A: T1WI; B: T2WI; C: Arterial phase; D: Venous phase; E: Delayed phase; F: Coronal view). G: The tumor was located in the upper left abdomen, highlighting the body surface. H: Showing the variation in the trend of CTR both preoperatively and postoperatively. POD (postoperative day).

as a sarcoma. Abdominal exploration was conducted, following which intraoperative examination showed that the base of the tumor was located in the diaphragm oppressing the left liver lobe. The base of the tumor, together with that partially surrounding the diaphragm, pericardium base, and the left lateral hepatic segment, was resected. The defect in the diaphragm and pericardium was repaired by patching (as guided by video), and the thoracic close drainage and abdominal drainage were placed following the operation (Figure 2).

After the operation, the patient was treated with antibiotics, expectorant, fluid complement, and nutritional support. The cardiothoracic ratio (CTR) changed from 0.66 to 0.60 (Figure 1D). The temporary rise in CTR was due to be free from the pericardium. He was discharged on post-operative day (POD) 14. The postoperative pathological report showed a giant solitary fibrous tumor (SFT). The following proteins were expressed in the cancer cells: CD34 (-), CD99 (-), NF (-), bc12 (+), Des (-), Act (-), S100 (-), Ki67 (+) 1%, β -catenin (+) (Figure 3).

Giant solitary fibrous tumor of the diaphragm

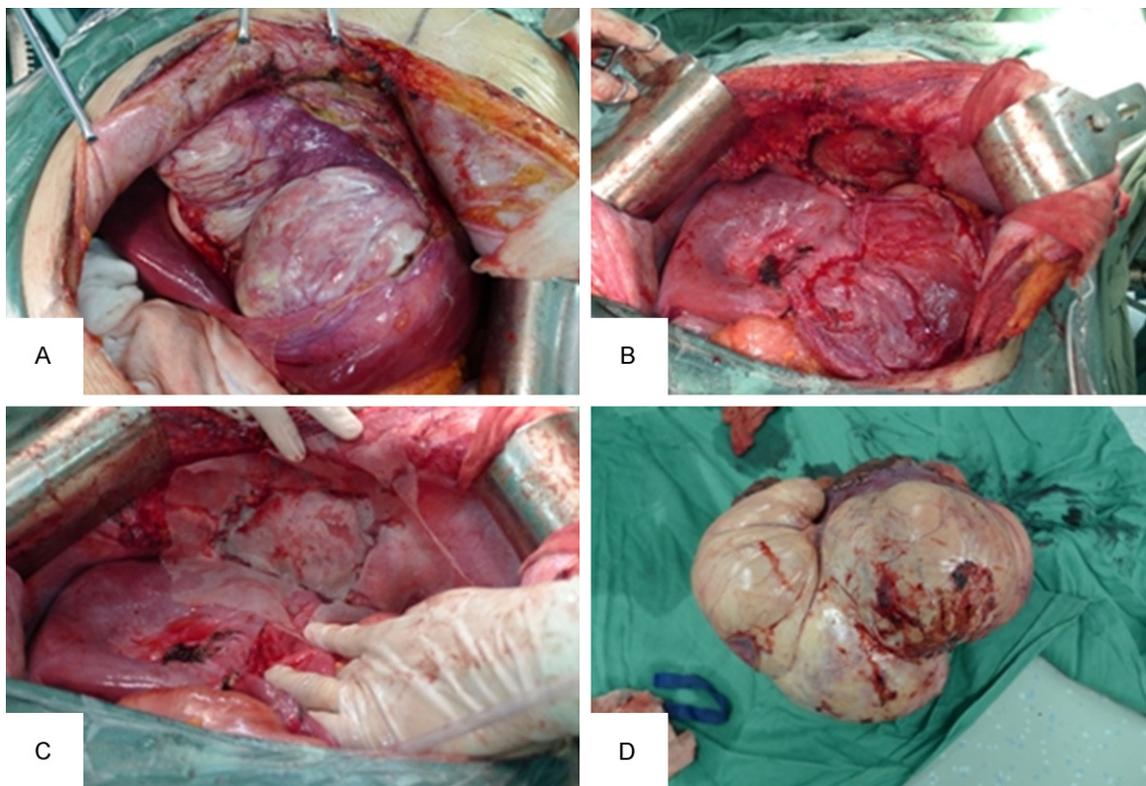


Figure 2. A. The tumor was located in the left upper quadrant, was lobulated, and compressed against the surrounding organs. B. The tumor was removed completely, including part of the diaphragm, showing that the left liver is pressed as a paper. C. A patch is used to repair the defect of the diaphragm. D. The tumor was well-defined, and determined to be a hard mass, with dimensions of 24 × 20 × 14 cm, weighing 3230 gm.

Discussion

SFT is a rare spindle cell tumor that arises from the visceral pleura according to the earliest study [2]. Later, SFT was identified in various locations outside of the thoracic cavity, such as the meninges [3, 4], orbit [5], nasal cavity [6], salivary gland [7], parapharyngeal space [8] and the paranasal sinuses [9]. SFT rooting in the diaphragm is very rare clinically. To the best of our knowledge, only four cases have been reported in the literature. In 1997, Norton [10] first reported an SFT of the diaphragm, occurring in a 60 year old woman with a two year history of respiratory symptoms. The lesion was initially misdiagnosed as an elevated left hemidiaphragm and confirmed through operative exploratory findings and histological appearance. In 2010, Ota [11] reported a case of diaphragmatic FTP that was successfully diagnosed by ultrasonography (US). The US images revealed a well-circumscribed mass with the feeding arteries leading from the diaphragm. Subsequently, this finding was confirmed histo-

pathologically after resection. In 2010, Lan [12] described a 32-year-old female smoker that presented with clinical features typical for gallstones. A chest CT demonstrated a large mass in intimate contact with the left hemidiaphragm and a laparoscopy was performed to confirm that the mass was associated with the diaphragm itself. Thus, a full thickness excision of the hemidiaphragm with the tumor contained within its leaves was completed. The hemidiaphragm was reconstructed using Gorx-Tex. In 2012, Kita [13] reported a case of SFT from the diaphragmatic pleura. A 71-year-old female was admitted to hospital because of an imaging study that revealed a mass on her right diaphragm. Surgical procedure was performed to remove the tumor, which was diagnosed as SFT with malignant potential through pathological examination and immune-histochemical findings.

SFT rooting in the diaphragm had no specific clinical manifestation [11-13]. The patients felt chest pain and tightness when the tumor

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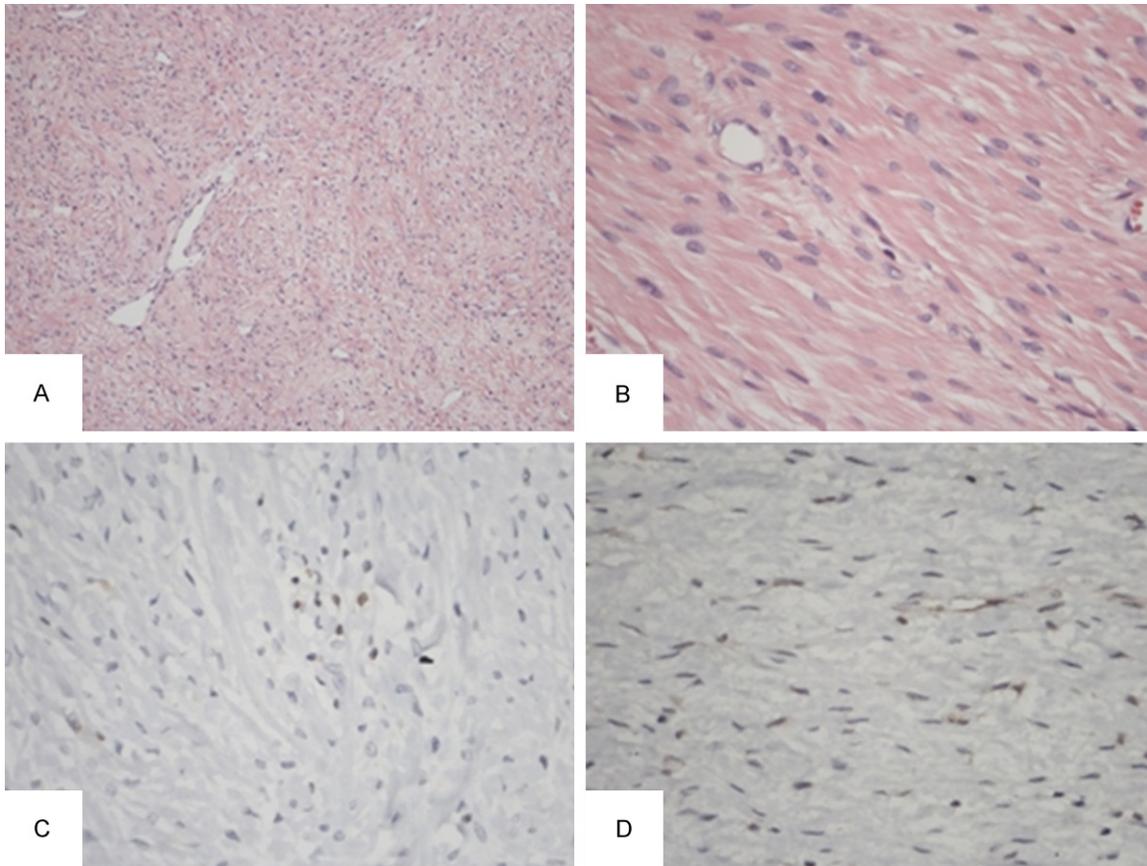


Figure 3. Microscopic examination showed proliferating spindle cells with a bundle or flow pattern (as determined by hematoxylin and eosin staining). A, B. Clusters of spindle cells are readily identifiable. No nuclear atypia, mitotic activity or necrosis were evident ($\times 100$ and $\times 400$ magnification). C. Immunohistochemical examination showing that the tissue was β -catenin -positive. D. Immunohistochemical examination showing that the tissue was bcl-2-positive.

oppressed the chest cavity and abdominal pain and distension when the tumor oppressed the abdominal cavity. Therefore, the clinical manifestation was closely related to the size of the tumor.

SFT does not display any tumor markers and the diagnosis relies on radiological examinations such as type-B ultrasonic, CT and MRI scans [14]. The tumors should be considered originating from the diaphragm when the type-B ultrasonic scanning shows the largest diameter in the diaphragm or extrahepatic area. Hideki [11] described a successful diagnosis of diaphragmatic SFT by preoperative ultrasonography. The mass was shown to be a well-circumscribed mass in the left thoracic cavity with the feeding arteries leading from the diaphragm. The specimen that was obtained from a solid component by US-guided percutaneous core needle-biopsy demonstrated bland-looking spindle cells alternating with numerous

bundles of small collagen fibers. The tumor cells showed immune-crossreactivity for vimentin and CD34 but not for keratin, and a diagnosis of diaphragmatic SFT was made.

Thus, US may prove to be a useful method for preoperative diagnosis of diaphragmatic SFT. CT and MRI can show the relationship between the tumor and diaphragm, and the tumors are usually detected as a sessile mass in the diaphragm [11]. However, the final diagnosis relies on surgical research and pathological examination [10-13]. The pathological features of SFT show that the tumor was comprised of spindle or short spindle cells and varying quantities of vascular tissue. Immunohistochemical examination revealed diffuse reactivity for CD34 and vimentin and positive expression of bcl-2 (80-100%) and CD99 (75-100%). However, the specimens did not express EMA or S-100 proteins [15]. This typical presentation of characteristics was evident in the present case as

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well. The positive expression for bcl-2 and negative expression of S-100 protein was observed (Figure 3).

SFT rooting in the diaphragm is easy to be misdiagnosed and to miss diagnosis. This tumor is likely to be diagnosed as deriving from the lung when growing in the chest. The right diaphragm mostly covers the surface of the liver, so this tumor is likely to be diagnosed as deriving from the liver when growing in the abdominal cavity and oppressing the liver. Homologous, this tumor is also easy to be diagnosed as deriving from the stomach, pancreas, spleen and posterior peritoneum.

Tumor rooting in the diaphragm cannot be confirmed as benign or malignant preoperatively; complete en bloc resection is the optimal treatment [10-13]. Benign tumor and boundaries clear of the malignancy should be removed including near the normal diaphragm and peritoneum. Once the tumor infringes the chest wall, the infringed chest wall should be removed. If the defect of the diaphragm is too large to suture, artificial materials such as fascia, polyester fabrics, and marlex can be used to repair the defect.

In the present case, we removed the tumor completely including infringed diaphragm and a wool patch was used to repair the defect of the diaphragm. The tumor was demonstrated as being SFT, so neither radiotherapy nor chemotherapy was required. SFT rooting in the diaphragm has a good prognosis. This patient had an uneventful postoperative course and he is currently well with no clinical signs of recurrence four months after his operation.

In conclusion, SFT rooting in the diaphragm is an orphan disease with no special clinical manifestation and preoperative diagnosis depends on imaging-based examinations. Surgery is the priority first choice treatment. The prognosis is considered well if the tumor is removed completely.

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Disclosure of conflict of interest

None.

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