

Case Report

Giant cardiac solitary fibrous tumor with 4 years follow-up: a case report and review of literature

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Abstract: Cardiac solitary fibrous tumors (SFTs) are extremely rare. A 22-year old young woman presented with dyspnea and heart failure was admitted to our hospital. Echocardiography and Computed tomography (CT) scan revealed a huge intrapericardial cystic mass (18 cm maximum diameter) with a massive pericardial effusion. The tumor was surgically resected, grayish white cystic neoplasm diagnosed with giant cardiac SFT by histology while absent of malignant features. The patient is symptom-free 4 years after surgery and undergoing close follow-up at regular intervals. This case report may provide valuable informational reference resource for the diagnosis and management of cardiac SFT.

Keywords: Solitary fibrous tumor, SFT, Cardiac neoplasm

Introduction

Solitary fibrous tumor is a mesenchymal tumor of fibroblastic type, which can affect any region of the body, but exceedingly rare presented in the heart, only a dozen cases reported in the literature up to date [1-3]. Primary cardiac tumors may cause severe systemic symptoms including heart failure [1]. Although most SFTs are benign, recurrence and aggressive malignancy subtype been reported [4-6]. Preoperative diagnosis of cardiac SFT is challenging because it is very difficult to define on the basis of cardiac imaging. This case report aims to provide information on clinical experience, diagnostic imaging outcomes and histopathological features, and treatment, (and) prognosis of cardiac SFT. Written informed consent was obtained from the patient.

Case report

The patient was a 22-year-old female, who was hospitalized with dyspnea and heart failure, with edema of lower extremities for 1 month before admission. Echocardiography showed giant cardiac cystic mass, Computed tomography (CT) scan revealed a huge intrapericardial cystic mass (18 cm maximum diameter) with a

low density massive and pericardial effusion, and CT enhanced scanning did not found enhancement region, while obviously sign of heart compression, with decreased volume (**Figure 1A, 1B**).

The pericardium tumor was successfully surgically resected, giant grayish white cystic neoplasm (20 cm maximum diameter) diagnosed with giant cardiac SFT by histology while absent of malignant features (**Figure 1C, 1D**). The postoperative course was stable, the patient had completely free of any symptom, was dismissed from the hospital after the tenth postoperative day.

Pathological findings

Macroscopically, multiple cyst-form tumor tissue had maximum size of 12 cm in diameter, 1 cm thickness grayish white cystic tissue, inner wall were smooth, bright pale color soft texture, no major hemorrhage or necrotic feature were present (**Figure 1C, 1D**).

Histological examination: Tumor composed spindle-shaped cells, patternless architecture of alternating hypo- and hyper cellular areas, cells with clear boundary, cyst wall is mainly consti-

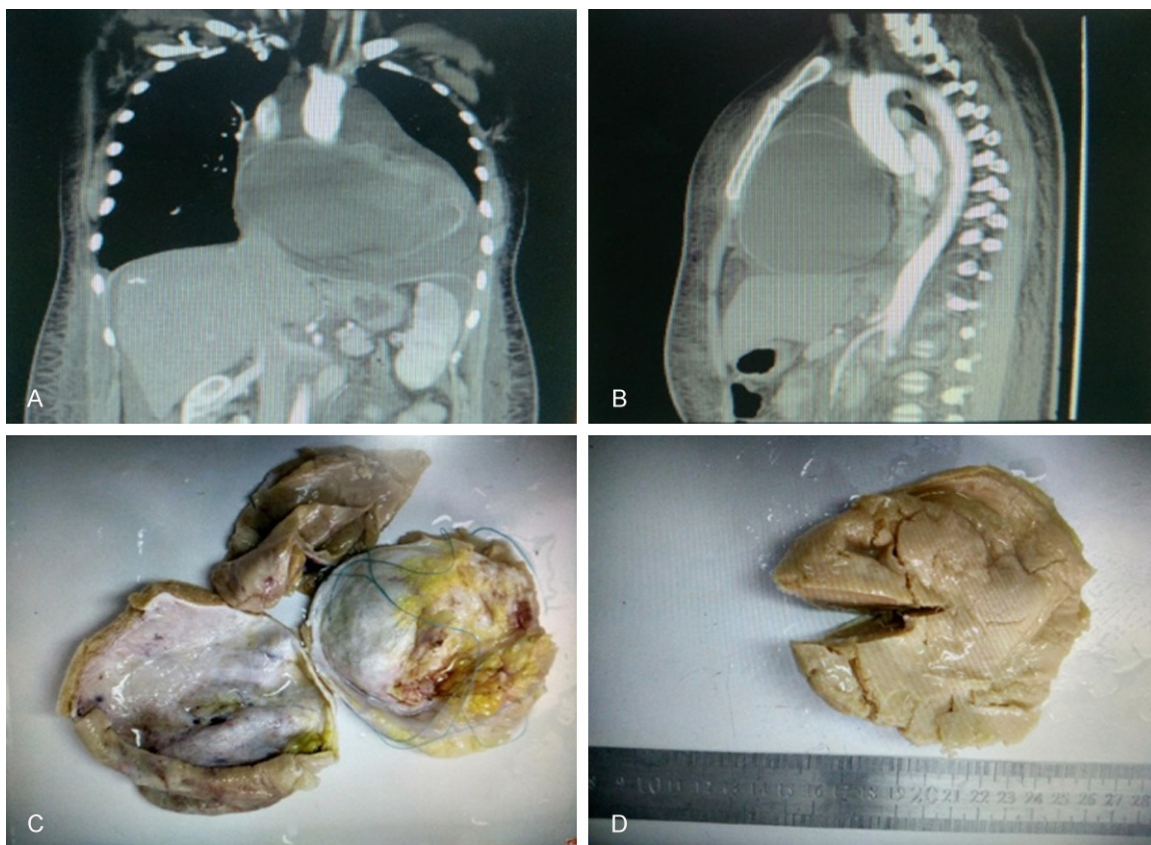


Figure 1. A, B: Computed tomography (CT) images, huge intrapericardial cystic mass, the maximum diameter was about 18 cm; C, D: Pathological findings images, giant grayish white cystic neoplasm, inner wall were smooth, bright pale color soft texture, no major hemorrhage or necrotic feature were present.

tuted by the tumor tissue, collagenous fibrous tissue coating the inner surface, no obvious epithelial lining, no necrosis tissue; nuclear mitoses were rare; part of the region of with varying proportions of collagen fibers component.

Immunohistochemistry results: It shown positive staining for CD34 (**Figure 2B**), Bcl-2, CD99, vimentin (**Figure 2C**), smooth muscle actin (SMA) (**Figure 2D**), and STAT6, EGR1 nuclear positivity (**Figure 2E, 2F**), while focally positive for desmin, S-100 protein. The Ki-67 positive index is 10% (**Figure 2G**), cytokeratin (CK) protein, epithelial membrane antigen (EMA), CD31, CD117 (**Figure 2H**), Dog1, calretinin (**Figure 2I**), inhibin are negative.

Pathological diagnosis: Giant cardiac cystic solitary fibrous tumor.

Follow up: At 4 years of follow-up, the patient was free of symptoms and without sign of tumor recurrence.

Discussion

Solitary fibrous tumor (SFT) is a mesenchymal tumor of fibroblastic type occurs predominantly in middle-aged adults with equal gender distribution. It can affect virtually almost in all anatomic sites, most common sites for pleura, head and neck, eyes, soft tissue and abdomen, extremities, cerebral meninges, while is extremely rare in the heart [1, 2]. In this case, the primary tumor site is in the right ventricular outflow tract. Systemic review the literature, only a dozen cardiac SFT cases in total reported so far. It's occurred mainly in the elder, pericardial location, atrial, ventricular, left ventricular outflow tract and right ventricle wall; tumor size (from 3 to 20 cm); most are benign tumor. In this case, as the giant tumor was located in the right ventricular outflow tract, heart was in severe compression, give rise to clinical symptom/sign of dyspnea and heart failure.

SFT is rare spindle cell tumor, originated in the pleura, may originate from pluripotent mesen-

Cardiac solitary fibrous tumor

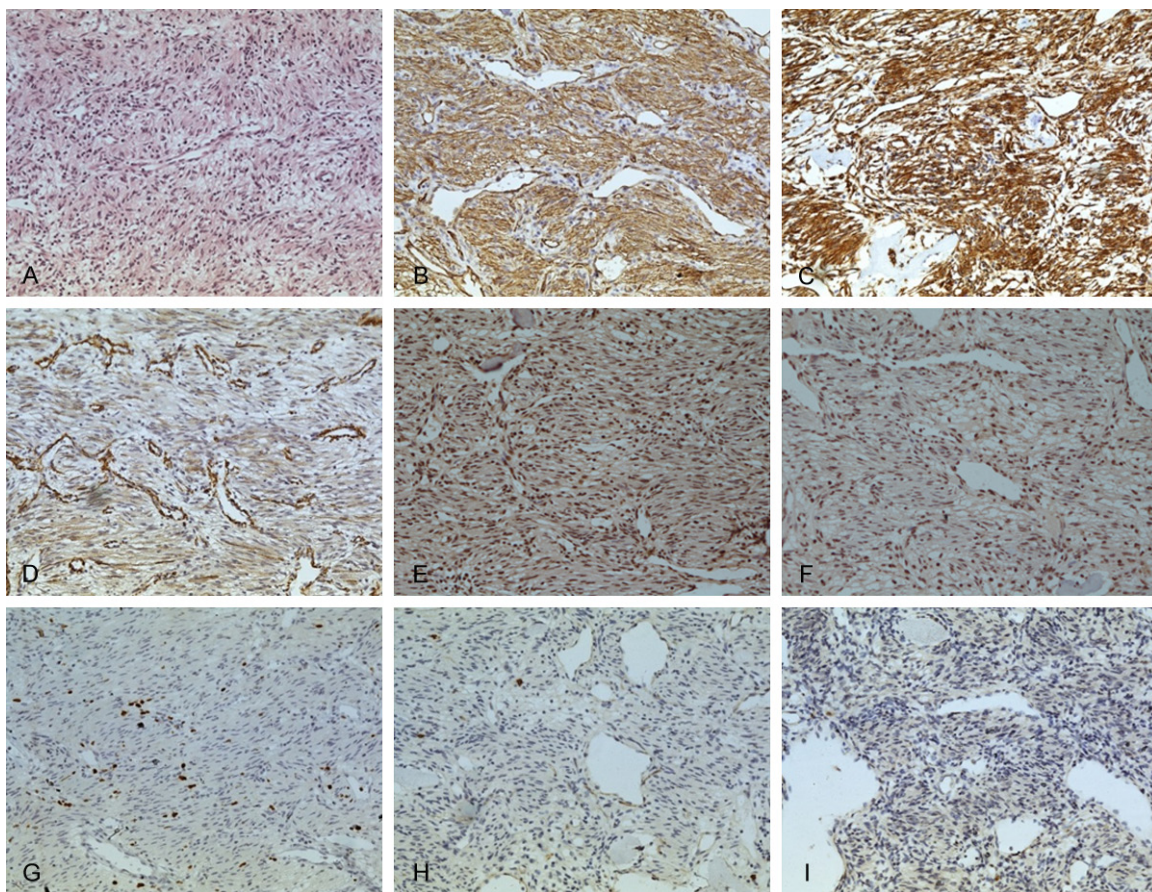


Figure 2. The IHC staining of tumor tissue in the heart were observed by Photomicrograph (magnification, $\times 200$). A: H&E stained from the patient; B-F: Immunohistochemical positive staining for CD34, vimentin, SMA, STAT6 and EGR1; G: Ki-67 positive index 10%; H, I: Negative staining for CD117 and calretinin.

chymal cells potential to differentiate into fibroblasts and myofibroblasts direction.

SFT has two common microscopic patterns. The first is described as a pattern of bland, wavy appearing spindle cells in a collagenized stroma. With alternating hypo- and hypercellular areas and a prominent branching vasculature. Because of this variable histological structure SFTs might be mistaken for other tumors such as hemangiocytoomas or fibromas. SFT has a variety of growth patterns, forms and manifestations. Due to complexity and diversity of morphology display, easily confused with some other spindle shaped tumors, especially in the cardiac site with more primary pericardium and epicardium tumor been reported recently. However, recent immunohistochemical tests (CD34, bcl-2 oncoprotein, CD99) allow separating SFTs from tumors with similar histomorphological characteristics [7].

Most SFTs are benign while its biological behavior is sometimes difficult to define. As atypical and malignant variants account small percentage of SFTs (10-20%), behave in a more aggressive way, tend to recur and metastasize. with local recurrence and/or distant metastasis for which systemic therapy (chemotherapy or targeted treatment with e.g. sunitinib) can be given. Prediction of behavior is difficult, with tumor size above 15 cm, positive surgical margins, tumor site and high mitotic count ($>4/10$ high power fields, HPF) being the most useful indicators for malignancy. The atypical and malignant variants, the histologic features of malignancy include increased mitotic activity, nuclear pleomorphism, and necrosis.

Most cardiac SFT are benign, so surgical resection is curative in most cases. Due to nature rarity of cardiac SFT, its biological behavior is difficult to predict, and tumor may recur after

surgery, there still no target therapy so far. So a thorough regular follow-up, if necessary, long-term should performed. For this patient obtain symptom-free 4 years after surgery, and we will continue to examine her at regular intervals.

Recent molecular genetics findings identified NAB2-STAT6 as the most frequently genetic alteration in SFTs [8-10]. STAT6 immunohistochemistry is a practicable powerful tool in diagnosing SFTs. This case shown immunohistochemistry positivity of STAT6, EGR1 activation support the notion they may as useful new markers in diagnosing SFTs. Also, the identification of the NAB2-STAT6 fusion gene can provide important diagnostic information, even though currently therapeutic options for more advanced or aggressive tumors are scarce and no standard modality for metastasized SFT.

In conclusion, giant cardiac SFT is exceptionally rare. Currently, histology and immunohistochemistry diagnosis are the standard and pivotal for diagnosis and prognosis. For the rarity and unpredictable characteristic of some SFTs, close follow-up with novel biomarker and targeting strategy are urgently needed for the better management of such unmet clinical challenge.

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Disclosure of conflict of interest

None.

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