

Case Report

Giant Solitary Fibrous Tumor of the Ascending Aortic Wall Causing Reversible Heart Failure: A Case Report and Review of the Literature

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Abstract

A 56-year-old woman was admitted to our hospital with a 2-week history of chest tightness and fatigue, and an echocardiogram revealed a massive polyserous cavity effusion. A massive (13.5 cm maximum diameter) intrapericardial mass was discovered using computed tomography (CT) and cardiovascular magnetic resonance imaging (MRI) in the ascending aortic wall. A pericardial biopsy was performed and diagnosed as a solitary fibrous tumor (SFT). After successful mass resection, an immunohistochemical test was positive for CD34, STAT-6, CD34, and Bcl2, which indicates a giant benign solitary fibrous tumor of the ascending aortic wall. After three years of follow-up, the patient is symptom-free, and histological indications of malignancy were absent. A giant benign solitary fibrous tumor is extremely rare in the heart, especially from the ascending aorta wall, and experience with this tumor location is limited, so close follow-up at regular intervals is considered necessary. We present this case, followed by a literature review on SFTs involving the heart and management approaches.

Keywords

cardiac tumors; solitary fibrous tumor; ascending aortic; heart failure

Introduction

Cardiac tumors are particularly rare, with an incidence of about 0.0017%–0.33% [1], but they are often accompanied by cardiac and systemic complications such as heart failure. Cardiovascular tumors are frequently diagnosed and treated using multidisciplinary combined therapy in cardiovascular medicine, cardiovascular surgery, radiology, and pathology. A Solitary Fibrous Tumor (SFT) is a rare type of spindle cell tumor of mesenchymal origin. Most cases are solitary pleural fibrous tumors. It has since been found that they can occur anywhere in the

body and are called extrapleural SFT, but very rarely in the heart [2,3]. In particular, large SFT in the ascending aortic wall is extremely rare [2,4]. Cardiac imaging examination is the primary auxiliary examination of cardiac tumors, but some cardiac malignant tumors cannot be excluded by imaging, so histopathology is the gold standard of diagnosis. Echocardiography is important for routine screening of cardiac tumors and long-term postoperative follow-up, but cardiac computed tomography (CT) and magnetic resonance imaging (MRI) detection can further understand the size, location, adjacent relationship with surrounding tissues, and lesion signal characteristics of the heart [5]. In this paper, we report a case of a massive benign SFT originating from the ascending aorta. The case was diagnosed by echocardiography, cardiac CT, and cardiac MRI and confirmed by histopathology of the mass. After surgical resection of the tumor, the patient's cardiac function was improved, and clinical symptoms were relieved. After 3 years of follow-up, the patient's general condition was satisfactory, and there was no recurrence. Up until 2022, few cases of SFTs with cardiac involvement had been reported.

Case Report

Clinical and Radiographic Presentation

The 56-year-old female patient was admitted to the Second Affiliated Hospital of Zhengzhou University on November 19, 2019, with a 2-week history of chest tightness and fatigue. The patient's vital signs showed a blood pressure of 88/68 mmHg, a heart rate of 102 bpm, a respiratory rate of 29 breaths per minute, and a temperature of 36.2 °C. Physical examination revealed jugular venous engorgement and peripheral edema. There was decreased respiratory sounds in both lower lungs, increased bilateral cardiac borders, distant heart sounds, no murmurs, and pericardial friction sounds. The patient's abdomen was soft and slightly distended; positive abdominal mobility was expressed. The patient had been healthy in the past and had no abnormal neurological findings. Laboratory reports indicated that: liver function: ALT 1239 U/L (reference range 9.0–50.0 U/L), AST 1403 U/L (reference range 15.0–40.0

U/L); cardiac function markers, tumor markers, and other laboratory reports showed no obvious abnormality. Ultrasound showed reduced left ventricular diastolic function, reduced left ventricular systolic function, a right-sided solid space-occupying focus at the bottom of the pericardium, and multiple serous effusions.

Chest and abdomen enhanced CT scan showed a 65.7 mm × 102.3 mm mass with heterogenous enhancement in the right anterior mediastinum (Fig. 1A,B) and fluid density shadow in the pericardium, bilateral thorax, and pelvic cavity. Chest MRI revealed an irregular mass mixed with slightly longer T1 and T2 signal shadows visible in the pericardial cavity of the right anterior area of the heart, along with small cystic long T1 and T2 signal shadows seen inside. Diffusion-weighted imaging (DWI) showed a slightly high signal, the Apparent Diffusion Coefficient (ADC) image shows a low signal locally, and after enhancement, obvious uneven enhancement can be seen, with the size of about 67 mm × 99 mm × 96 mm. The boundary with the adjacent right atrium was not clear (Fig. 1C,D), and the right atrium showed a compression change. Considering the position of the solid space-occupying lesion within the pericardium, the origin of the mesenchymal tissue was possible.

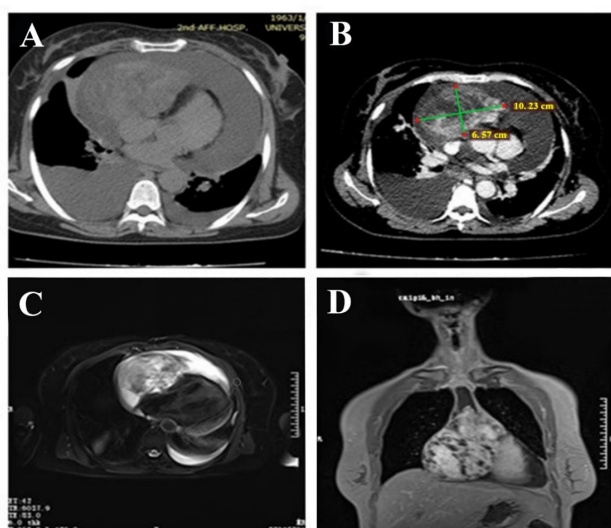


Fig. 1. CT and MRI imaging examination. (A,B) Chest computed tomography (CT) showed a mass with mixed density shadow in the right anterior mediastinum, uneven enhancement on enhanced scan. (C,D) Chest magnetic resonance imaging (MRI) showed an irregular mass of signal shadow can be seen in the right anterior area of the heart in the pericardium. After enhancement, obviously uneven enhancement can be seen, and the boundary with the adjacent right atrium is not clear.

Pathological Findings

Ultrasound-guided puncture biopsy was performed on the pericardial space-occupying cells. The biopsy specimen was 17 mm × 1 mm in size, and gray and white in color. Microscopically, the tumor cells were spindle-shaped cells in a mild form, arranged in bundles and braided shapes, with moderate atypia and a rare mitotic image. The immunohistochemical test was positive for CD34, STAT-6, CD34, and Bcl2. Diagnosis: A space-occupying lesion in the pericardium, consistent with a solitary fibrous tumor in the ascending aortic arch.

The mass was surgically resected. During the operation, it was found that the tumor was red and white in color, mixed with cystic and fibrous tissue, and had both medium softness and hardness in texture. The surface was smooth, the envelope was complete, no adhesion to the heart or pericardium was present, and the boundary was clear. The tumor was pedicled from the wall of the ascending aorta and covered the heart in an “umbrella” form (Fig. 2A–C). The specimens were fixed in 10% neutral formalin, paraffin-embedded, stained with hematoxylin and eosin, and examined under a light microscope. Immunohistochemical streptavidin-peroxidase staining was performed at the same time.

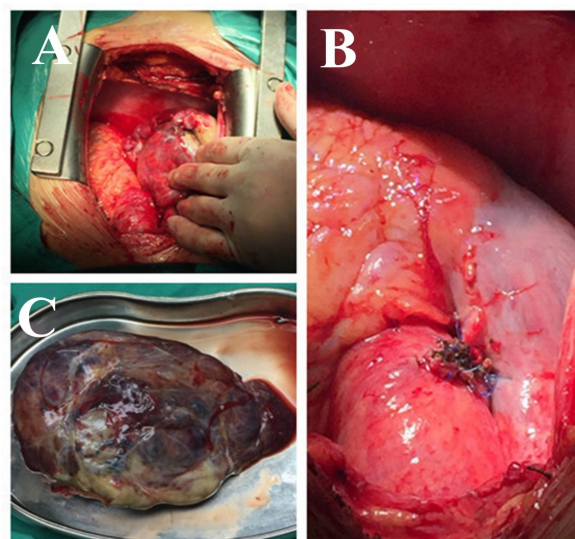


Fig. 2. Masses removed from the ascending aorta. (A–C) An intraoperative tumor was found on the wall of the ascending aorta covering the heart in the form of an “umbrella”. There was no adhesion between the tumor and the heart or pericardium, and the boundary was clear. The tumor was completely extracted.

The lesion was an encapsulated tumor 135 mm × 120 mm × 40 mm in size and grayish-red (Fig. 3A). The capsule was complete. Histologically, the tumor consisted of spindle-shaped cells that were arranged in a braided shape. The cells were moderately heteromorphic, and mitosis was

rare. The immunohistochemical test was positive for vimentin, CD34, CD99, STAT-6, and Bcl2 (Fig. 3B–D). The final pathological diagnosis was an ascending aortic wall vascular spindle cell tumor, with immunohistochemistry consistent with an isolated fibrous tumor, based on the findings described above. Clinical diagnosis: benign SFT in the ascending aortic wall.

The patient was closely monitored. Three years after surgery, the patient is now in good condition, without cardi-ological symptoms, and there has been no tumor recurrence.

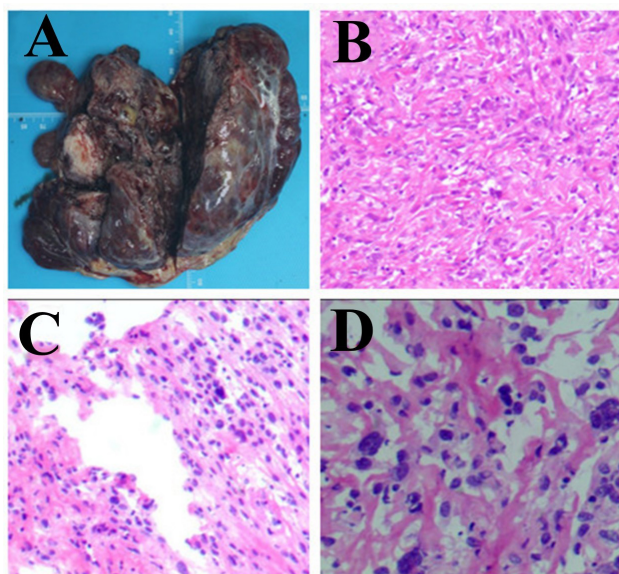


Fig. 3. Histopathological examination of the tumor. (A) The mass following complete resection. (B–D) Histological staining revealed spindle-shaped cells with abundant collagen.

Discussion

SFT was first reported by Klemperer and Rabin in 1931 and is defined as a tumor originating in the pleura. SFT usually occurs in the pleura, but studies have found a diffuse distribution of CD34-positive mesenchymal cells in human connective tissues, suggesting that SFT can occur in all anatomical sites of the body. In scientific literature, there have been nearly 20 previous case reports of a primary cardiac SFT [1–19] and only 2 case reports of ascending aorta and pulmonary trunk SFT; however, the origin of ascending aorta alone has not been reported.

Some features of these case reports were summarized in Table 1 (Ref. [1–20]). With limited data, we found that cardiac SFT can occur at any age, but mainly in middle-aged individuals, with no significant gender difference. Three of these cases were malignant SFT, 15 were benign, and 2 were unidentified. The majority of clinical manifestations are slow local growth of painless masses, and with

tumor growth often appearing at the corresponding site of compression symptoms, the clinical manifestations are primarily related to the location and size of the mass. Most patients present with symptoms of heart failure due to diastolic insufficiency caused by large tumors. Five patients had no clinical symptoms and were accidentally found to have painless masses with clear peripherals during physical examination. Therefore, selecting an appropriate imaging examination is of great significance for the early detection of lesions.

Radiologically, SFTs are variable and generally non-specific. In the case of heart SFT, enlargement of the heart shadow can usually be found on chest X-ray; echocardiogram can indicate space-occupying lesions in the pericardium and pericardial effusion. The enhancement is obvious in the densely populated area of tumor cells, while relatively weak in the sparse area of cells and the hyaloid area of collagen fiber bundles form the “map”-like heterogenous enhancement. When the density (or signal) is uneven on imaging, cystic degeneration, necrosis, calcification, and bleeding are considered possible causes. There is no enhancement of the cystic necrosis area; the irregular necrosis area is relatively common in malignant tumors, and malignant lesions are suspected when uneven enhancement occurs in the center of the lesion.

Cardiac MRI can further clarify the shape, location, and relationship between the tumor and surrounding tissues, thus elucidating the nature of the tumor. Generally, SFT on T1WI presents low-to-intermediate signal intensity signals, while T2WI signals are non-homogeneous. The intensity of T2WI signals decreases with the increase of collagen fiber content, while T2WI signals present high signals when cells are dense and blood vessels are abundant, which is of great diagnostic value. The blood supply of the tumor is abundant. Empty blood vessels can be seen in a plain scan, and significantly enhanced blood vessels can be seen in and/or around the tumor after enhancement.

The final diagnosis of SFT depends on pathology, especially immunohistochemical staining. Microscopically, a large number of spindle cells are arranged in bundles or braid shapes under the collagen matrix, and the cells had no obvious atypia. The collagen fiber bundles are arranged in parallel, and the interstitial blood vessels were abundant. Malignant SFT is characterized by abundant cells, increased cell density, and high cell atypia, with high mitotic activity (>4 cells per 10 highest possible frequency), and a similar morphology to fibrosarcoma and pleomorphic undifferentiated sarcoma with frequent secondary hemorrhagic necrosis, and invasive growth at the edges [20]. The maximum tumor diameter is usually more than 100 mm. Immunohistochemical staining mostly indicated positive CD34 and vimentin, and other positive markers included CD99 and Bcl2 [21]. The most important immunophenotypic feature supporting SFT was concurrent positive CD34 and Bcl2.

Table 1. Summary of case reports of primary cardiac solitary fibrous tumor (SFT).

NO./Reference	Year	Age (y)/Sex	Symptoms	Location of primary tumor	Tumor size	Tumor type	Positive immuno-histochemical markers	Follow-up
1. El-Naggar [6]	1989	56 y/F	NA	pericardium	130 mm max diameter	Benign	NA	202 months
2. Bortolotti, U [2]	1992	60 y/M	fatigue, chest discomfort, and increasing dyspnea	AAo, PT	140 × 70 mm	Benign	Vimentin	9 months
3. Segawa, D [7]	1995	50 y/F	dyspnea and palpitation	RV	125 × 105 × 80 mm	NA	Vimentin	19 months
4. Flemming, P [5]	1996	53y/F	asymptomatic	LV	NA	NA	CD34, Vimentin	died
5. Andreani, SM [8]	1998	60 y/M	exertional dyspnea, irregular lower extremities and face edemas	pericardium	200 × 180 mm	Benign	NA	48 months
6. Corgnati, G [4]	2004	30 y/M	Asthenia,	AAo, PT	120 mm	Benign	NA	18 months
7. Bothe, W [9]	2005	39 y/F	palpitations	RA	44 × 45 × 55 mm	Benign	CD34, Vimentin	12 months
8. Croti, UA [10]	2008	5 m/M	asymptomatic	LA	30 mm ²	Benign	CD34	Died after 6 months
9. Zhao, XG [11]	2012	55 y/M	chest distress and shortness of breath	RA	150 × 100 × 80mm	Malignant	CD34	Died after surgery
10. Tamenishi, A [12]	2013	30 y/F	syncope	ltPA	90 × 74 × 51 mm	Benign	CD34	72 months
11. Taguchi, S [13]	2013	49 y/F	asymptomatic	LV	150 × 90 × 55 mm	Benign	CD34, Vimentin, CD99	NA
12. Bianchi, G [1]	2013	68 y/F	dyspnea and fatigue	LV	170 mm max diameter	Benign	CD34, Vimentin, Bcl2	12 months
13. Czimbalmos. C [3]	2017	37 y/F	asymptomatic	PT	100 × 40 × 90 mm	Benign	CD34, Vimentin, CD99, STAT6	36 months
14. Kotani. S [14]	2018	68 y/M	asymptomatic	attached to a distal aortic	69 × 65 × 36 mm	Benign	CD34, (Bcl2), STAT6, and MIC2	6 months
15. Shao and Wang [15]	2019	51 y/F	exertional dyspnea, dry cough, and diffuse edema	Pericardium	46 × 68 × 75 mm	Benign	CD34, vimentin, CD99, Bcl2	NA
16. Zhang, L P [16]	2019	64 y/F	dyspnoea and abdominal distension	RA Pericardium	75 × 34 mm	Malignant	CD34, vimentin, Bcl2	NA
17. Fan, J [17]	2019	67 y/M	swelling and pain in the right leg	inferior vena cava (IVC)	50 × 40 × 30 mm	Benign	vimentin, CD99, Bcl2, CD34 and STAT6	6 months
18. Luo, R [18]	2020	70 y/F	oppression in chest	pulmonary artery	43.7 mm × 15.9 mm	Malignant	CD34(3+), d2-40(+), STAT6(±)	12 months
19. Chen, Lei [19]	2020	52 y/M	cough, chest pain, and dyspnea	middle mediastinal	85 × 95 mm	Benign	CD99, CD34, Bcl-2, and vimentin	69 months
20. Fain, Kristen [20]	2022	81 y/M	progressive dyspnea	left inferior mediastinum connect to pericardium	69 × 54 mm	Benign	Bcl2, CD34, and STAT 6	6 months
21. Our case	2023	56 y/F	Chest stiffness and fatigue	AAo	135 × 120 × 40 mm	Benign	CD34, Bcl2, CD99, STAT6	38 months

M, male; F, female; NA, no data available; AAo, ascending aorta; PT, pulmonary trunk; RV, right Ventricle; LV, left Ventricle; RA, right atrium; LA, left atrium; ltPA, left pulmonary artery; PA, left pulmonary artery.

Although the pathogenesis of isolated fibrous tumors in clinic has not been clearly identified, they may be closely related to the driving effect and changes in the *NAB2-STAT6* fusion gene, which is highly specific for SFT diagnosis. Immunohistochemical detection of STAT6 could be used to replace the above fusion gene detection, and more than 90% of the cases showed strong positive tumor nuclei [22]. In this case, Vimentin, Bcl2, and STAT-6 were diffusely positive; CD34 and CD99 were strongly positive; and Ki67 was highly expressed, which was consistent with CD34 negative MSFT.

Most SFT tumors, especially extrapleural tumors, are benign and treatable by full tumor resection, with the exception of those of mediastinal origin; nonetheless, there are 10–20% malignant lesions [23] that include cardiac SFT [13,24]. About 63% of malignant SFT relapsed, and 2% of morphologically benign SFT could also metastasize or transform into malignant SFT, which may be the manifestation of tumor dedifferentiation. This signifies that the morphology of SFT is not a complete predictor of prognosis, and the complete resection of the tumor and the guarantee of a negative resection margin are the key factors influencing the prognosis. Therefore, all patients should be closely followed-up for a long time. In the reported literature, chest radiographs and echocardiography are the most commonly used non-invasive methods for close follow-up. The CARE checklist was used when writing this case report (Supplementary Fig. 1).

Limitation

The limitation of our study was the inability to obtain follow-up CT scans, as the patient refused to undergo further imaging. However, we believe that the histological examination performed provided valuable insights into the nature of the patient's condition and the potential for future complications.

Conclusions

Massive cardiac SFT is extremely rare, especially in the ascending aorta wall. Tumors in the pericardium end up causing diastolic heart failure symptoms. Echocardiography, cardiac CT, and MRI can assist in diagnosis, but pathology and immunohistology will determine the final diagnosis. The prognosis is not always clear in cardiac SFT cases; hence all patients should be closely followed up for a long time.

Availability of Data and Materials

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions

PG designed this study and made a major contribution to the manuscript. SL, EMIA, XJ and LJ contributed to the data analysis and discussion. YW was involved in interpretation and revision. All authors contributed to editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work to take public responsibility for appropriate portions of the content and agreed to be accountable for all aspects of the work in ensuring that questions related to its accuracy or integrity.

Ethics Approval and Consent to Participate

This case report is a retrospective study that did not involve clinical trials and was exempt from the Ethics Committee of the second affiliated hospital of Zhengzhou university (Zhengzhou, China). We have got the patient's consent and all privacy information of the patient was protected.

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Conflict of Interest

The authors declare no conflict of interest.

Supplementary Material

Supplementary material associated with this article can be found, in the online version, at <https://doi.org/10.59958/hsf.5513>.

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