

# Pericardial malignant solitary fibrous tumour with right atrial invasion – a case report and literature review

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## Abstract

Solitary fibrous tumours are unusual neoplasms that develop from mesenchymal cells, usually originating from the pleura. A pericardial solitary fibrous tumour is an extremely rare occurrence. We report a 64-year-old woman who presented to the hospital with chief complaints of dyspnoea and abdominal distension. Echocardiography and enhanced computed tomography revealed an intrapericardial tumour with local invasion to the right atrium. Histopathological examination of a biopsy specimen showed a patternless distribution of spindle-shaped cells in a collagen stroma, with a high mitosis rate. Immunohistochemistry was positive for vimentin, CD34, and Bcl-2. The final diagnosis was a pericardial malignant solitary fibrous tumour with right atrial invasion. Surgical resection of the tumour was not performed because of its invasion into the myocardium. We systematically reviewed the literature on cardiac solitary fibrous tumours up to 2019.

## Keywords

Solitary fibrous tumour, pericardial localization, cardiac biopsy, histopathology, immunohistochemistry, pericardial tumour

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## Introduction

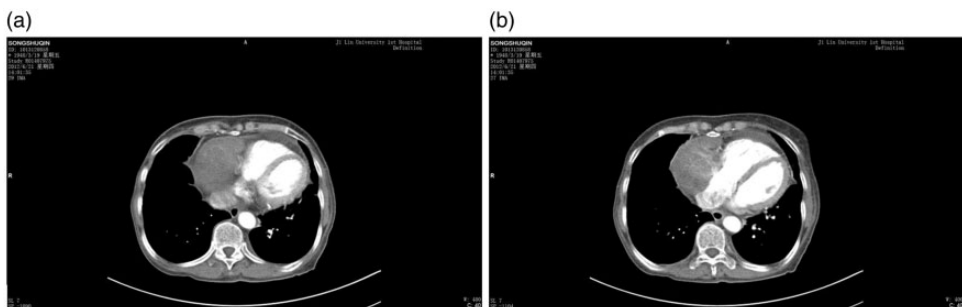
Solitary fibrous tumours (SFTs) are neoplasms derived from fibroblastic mesenchymal cells, usually originating from the pleura. Although extrapleural anatomic locations have been reported,<sup>1</sup> primary cardiac SFTs are extremely rare. The histological appearance of SFTs shows a spindle-shaped pattern and SFT cells also express several immunoreactive markers, including CD34, Bcl-2, CD99, and STAT6.<sup>2,3</sup> SFTs usually behave in a benign fashion and malignancy is rare, characterized by the presence of necrotic, polymorphic, mitotically active cells and aggressive behaviour.<sup>4,5</sup> Few cases of SFTs with cardiac involvement have been reported up to 2019.<sup>6–20</sup>

## Case report

A 64-year-old woman was admitted to hospital with a 1-month history of dyspnoea and abdominal distension. She had exertional dyspnoea that restricted her ability to perform daily activities. She also complained of abdominal distension, which was aggravated by food intake. She had a history of chronic bronchitis and smoking for the past 30 years. Physical examination showed a heart rate of 103 beats/minute, blood pressure 110/70 mmHg, respiratory rate 29 breaths/minute, mildly cyanosed lips, mildly distended jugular veins, and

decreased respiratory sounds, but no wheezes or crackles. Cardiac examination showed increased bilateral cardiac borders, a normal rhythm, no murmurs and pericardial friction sounds. Her abdomen was soft and the liver and spleen were not palpable. There was moderate pitting oedema of the bilateral lower limbs.

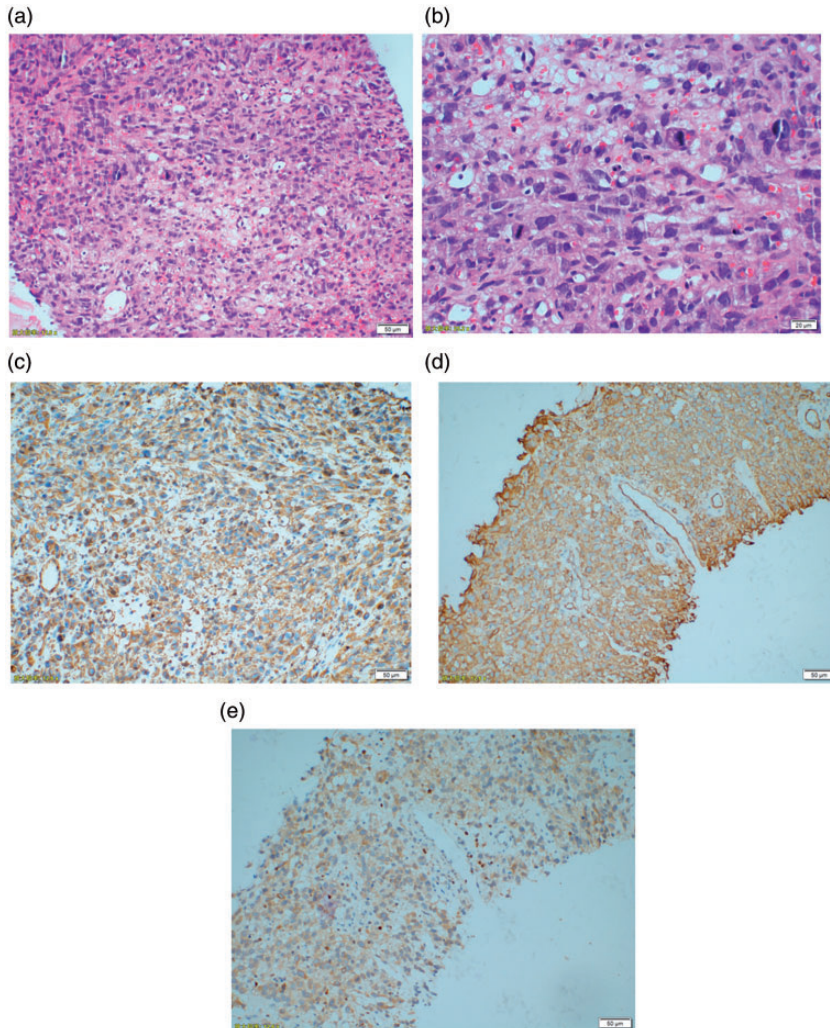
Laboratory reports showed a white blood cell count of  $12.58 \times 10^9/L$  (reference range  $3.5\text{--}9.5 \times 10^9/L$ ), neutrophils 84% (reference range, 40%–75%), N-terminal pro-brain natriuretic peptide 2550 pg/mL (reference range 0–125 pg/mL), and troponin 0.23 ng/mL (reference range 0–0.034 ng/mL). Tumour markers were neuron-specific enolase 21.4 ng/mL (reference range <25 ng/mL) and carbohydrate antigen 125 11.29 U/mL (reference range <35 U/mL). Echocardiography revealed an ejection fraction of 57%, moderate pericardial effusion, and a  $79 \times 37$  mm hyper-echoic mass in the pericardial sac, located in the visceral pericardium of the lateral wall of the right atrium. Enhanced computed tomography (CT) also revealed a  $75 \times 34$  mm intrapericardial mass. The tumour invaded the right atrial myocardium with inhomogeneous enhancement (Figure 1). The patient underwent pericardiocentesis and 300 mL of bloody pericardial effusion was sent to the laboratory for testing. The results showed exudative



**Figure 1.** Enhanced CT revealed pericardial effusion and a  $75 \times 34$  mm intrapericardial mass with inhomogeneous enhancement (a). The right atrial myocardium was invaded by the tumour (b).

pericardial effusion, adenosine deaminase 28.10 U/L (reference range 4–18 U/L), lactate dehydrogenase 767 U/L (reference range 120–250 U/L), and carcinoembryonic antigen 2.13 ng/mL (reference range <3.4 ng/mL). Tuberculosis-related tests on the pericardial effusion were negative. Ultrasound-guided cardiac biopsy was performed and histopathological examination

revealed a patternless distribution of oval and spindle-shaped cells in a collagen stroma, with mitosis >40 in 10 high-power fields. Immunohistochemistry was positive for vimentin, CD34, and Bcl-2 (Figure 2). Based on the above findings, the final diagnosis was pericardial malignant solitary fibrous tumour with right atrial invasion. Complete resection of the tumour was



**Figure 2.** Biopsy histopathology revealed a patternless distribution of oval and spindle-shaped cells in a collagen stroma, tumour cells with acidophilic chromosomes, and mitosis >40 in 10 high-power fields (a,b). Immunohistochemistry revealed intense cytoplasmic staining for vimentin (c), CD34 (d), and Bcl2 (e). HE: haematoxylin and eosin.

impossible because of its invasion into the myocardium. The patient refused chemotherapy and was discharged after pericardial drainage.

Written informed consent was obtained from the patient for publication of this case report.

## Discussion

Primary pericardial tumours are unusual, with a prevalence rate of only 0.001% to 0.007%.<sup>21</sup> Mesothelioma is the most common type of malignant pericardial tumour, but other types including sarcomas and lymphomas also occur.<sup>22</sup> SFTs are rare mesenchymal neoplasms that usually originate from the pleura, though extrapleural locations have been reported, including the abdomen, mediastinum, retroperitoneum, pelvis, meninges, extracranial tissues of the head and neck, and soft tissues.<sup>1</sup> Primary cardiac SFTs are extremely rare, with only 14 reported cases up to 2019<sup>7-20</sup> (Table 1). Except for one case of a 5-month-old child, cardiac SFTs usually occur in

middle-aged patients (30–68 years old). Extrapleural SFTs have a largely equal sex ratio, with a slight male predominance for the fat-forming variant (male-to-female ratio, 3:2).<sup>23</sup> However, the reported cases of cardiac SFTs included five men and nine women, suggesting a male-to-female ratio of about 1:2. Cardiac involvement in previous cases included the ascending aorta, pulmonary trunk, right atrium, left atrium, left ventricle, pulmonary artery, and pericardium (4 cases). Among the 14 cases, only two were diagnosed as malignant, 10 cases were benign, and two were unidentified, suggesting that malignant cardiac SFTs are relatively rare. Extrapleural SFTs are usually more aggressive than the pleural form, with an incidence of aggressive behaviour of 6% to 23%.<sup>4</sup> In the current case, biopsy histopathology showed a high mitotic index, and echocardiography and enhanced CT revealed that the pericardial tumour had invaded the right atrial myocardium, indicating clinically aggressive behaviour.

**Table 1.** Primary cardiac solitary fibrous tumours reported in the literature.

No.	Reference	Year	Age	Sex	Location	Malignancy	Positive immuno-histochemical markers
1	el-Naggar et al. <sup>7</sup>	1989	56	F	Pericardium	Benign	NA
2	Bortolotti et al. <sup>8</sup>	1992	60	M	AAo, PT	Benign	vimentin
3	Segawa et al. <sup>9</sup>	1995	50	F	RV	NA	vimentin
4	Flemming et al. <sup>10</sup>	1996	53	F	LV	NA	CD34, vimentin
5	Andreani et al. <sup>11</sup>	1998	60	M	Pericardium	Benign	NA
6	Corgnati et al. <sup>12</sup>	2004	30	M	AAo, PT	Benign	NA
7	Bothe et al. <sup>13</sup>	2005	39	F	RA	Benign	CD34, vimentin
8	Croti et al. <sup>14</sup>	2008	5m	M	LA	Benign	CD34
9	Zhao et al. <sup>15</sup>	2012	55	M	RA	Malignant	CD34
10	Taguchi et al. <sup>16</sup>	2013	49	F	LV	Malignant	CD34, vimentin, CD99
11	Bianchi et al. <sup>17</sup>	2013	68	F	LV	Benign	CD34, vimentin, Bcl2
12	Tamenishi et al. <sup>18</sup>	2013	30	F	ltPA	Benign	CD34
13	Czimbalmos et al. <sup>19</sup>	2017	37	F	PT	Benign	CD34, vimentin, CD99, STAT6
14	Shao and Wang <sup>20</sup>	2019	51	F	Pericardium	Benign	CD34, vimentin, CD99, Bcl2
15	This case	2019	64	F	RA Pericardium	Malignant	CD34, vimentin, Bcl2

M: male, F: female, NA: no data available, AAo: ascending aorta, PT: pulmonary trunk, RA: right atrium, LA: left atrium, ltPA: left pulmonary artery, LV: left ventricle, PA: pulmonary artery, m: months

The symptoms of cardiac SFTs may depend on the extent and anatomic location of the tumour. The diverse clinical manifestations usually include dyspnoea, fatigue, chest discomfort or distress, palpitations, syncope, and/or peripheral oedema, although some cases were asymptomatic. Cardiac SFTs may directly invade the surrounding structures and protrude into the pericardial cavity, potentially causing pericardial symptoms such as pericarditis or pericardial tamponade, as in our case.<sup>18</sup> The present patient presented with dyspnoea and abdominal distension and the pericardial effusion was bloody and exudative, suggesting that pericardial effusion or tamponade could be a clinical manifestation of cardiac SFTs. However, these symptoms were not typical and some previous patients were asymptomatic.<sup>19</sup>

The commonly used diagnostic modalities for cardiac tumours include echocardiography, cardiac CT, and magnetic resonance imaging (MRI).<sup>24</sup> Echocardiography can help to determine the shape and size of the intrapericardial mass and its relationship with peripheral structures, as well as the presence of pericardial effusion. Cardiac CT can provide information on the morphology, location, margins, and extent of the tumour and its invasion of the neighbouring myocardium and vessels. SFTs may show inhomogeneous contrast enhancement, as in our case, which indicates sufficient blood supply to the tumour. MRI may help to verify the characteristics of SFTs, such as vascularity, cystic degeneration, haemorrhage, and necrosis. SFTs usually show intermediate signal intensity on T1-weighted images and variable signal intensity on T2-weighted images, and contrast MRI may show septated, patchy, and inhomogeneous late gadolinium enhancement.<sup>19</sup>

The confirmed diagnosis of SFT relies on histopathological and immunohistochemical features. Characteristic histopathological

features include a patternless architecture of spindle-shaped cells in a variable collagen stroma. Malignant SFTs are usually characterized by the presence of infiltrative margins, pleomorphism, hypercellularity, a high mitotic index, and necrosis.<sup>23</sup> SFTs also normally express CD34, Bcl-2, and CD99.<sup>25</sup> Recent studies found that the pathogenesis of SFT was related to the presence of a NAB2-STAT6 fusion gene resulting from a paracentric inversion on chromosome 12q13.<sup>26</sup> Nuclear expression of STAT6 is thus the most specific marker for diagnosis of SFT.<sup>27</sup> Biopsy histopathology in the current case showed a typical patternless distribution of spindle-shaped cells with a high mitotic rate, suggesting a malignant nature, while immunochemistry revealed expression of vimentin, CD34, and Bcl-2, all of which conformed to the features of SFTs.

Treatment for thoracic SFTs involves surgical resection,<sup>28</sup> although surgery for patients with advanced or aggressive SFTs may be limited because of invasion of the myocardium or great vessels. Although some patients with cardiac SFTs were reported to live for more than 2 years after surgical resection of the tumour, one of the reported cases died immediately after surgery.<sup>15,19</sup> The shortage of case reports means that the prognosis of cardiac SFTs remains unclear, and a multi-centre registration study may be needed to increase the number of reported cases.

In conclusion, primary pericardial SFT is an extremely rare condition. The diagnosis of SFT is challenging and requires an integrated approach including clinical, histological, immunohistochemical, and molecular findings.

#### **Declaration of conflicting interest**

The authors declare that there is no conflict of interest.



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