A 68-year-old woman with a 2-year history of dyspnea and fatigue was admitted to our hospital with a massive pericardial effusion. Computed tomography and cardiovascular magnetic resonance imaging revealed a huge (17 cm maximum diameter) intrapericardial mass. After successful tumor resection, a giant solitary fibrous tumour of the epicardium was diagnosed by histology. Histologic features of malignancy were absent, and the patient is alive and well 1 year after the operation, undergoing close follow-up at regular intervals. Recurrences have been exceptionally reported in benign solitary fibrous tumors, and experience with this exceptionally rare and enigmatic cardiac tumor is lacking.

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Primary cardiac tumors are rare, their estimated frequency ranging from 0.0017% to 0.33%, but they may cause a variety of cardiac and systemic symptoms, including heart failure. Clinical and pathologic expertise is required for their diagnosis and management. Solitary fibrous tumor (SFT) is exceedingly rare in the heart, six cases having been described so far, including a giant SFT and a malignant SFT [1–6]. Cardiac imaging is the main diagnostic tool in cardiac masses; however, histology represents the gold standard for differential diagnosis because in some cases malignancy cannot be definitively excluded on the basis of cardiac imaging. Although echocardiography is the conventional screening and follow-up method for intracardiac masses, computed tomography (CT) and cardiovascular magnetic resonance imaging (CMRI) may add useful data about size and location, adjacent mediastinal structures, and signal characteristics [7]. In the present report, the complementary roles of cardiac imaging and histology are shown in an exceedingly rare cardiac giant SFT presenting with heart failure.

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A 68-year-old woman with a 2-year history of progressively worsening dyspnea and fatigue was admitted to our hospital with a diagnosis of massive pericardial effusion. By CT, a huge (15 × 17 cm) intrapericardial mass with vascularization from both the circumflex and the posterior descending coronary arteries was demonstrated (Fig 1A–C). CMRI also showed early contrast medium enhancement and late persistence in the inner core of the mass (Fig 2A, B). Preoperative echocardiography evidenced a normal biventricular function with dilated inferior vena cava and mild mitral valve regurgitation.

The tumor was blunt dissected from the diaphragmatic wall of the heart through a median sternotomy approach. On the obtuse margin of the heart, the tumor stalk was excised by use of electric cautery. To increase the likelihood of complete resection, the epicardial area corresponding to the tumor stalk was also cauterized. Arterial and venous feeders were ligated on the myocardial side, clipped on the tumor’s side, and then separated with scissors to avoid intraoperative bleeding. No hemodynamic instability occurred intraoperatively; thus, cardio-pulmonary bypass was not used. The postoperative course was uneventful, although during two-dimensional echocardiography before discharge a severe mitral regurgitation was found, possibly related to chronic distortion of the mitral valve apparatus. Because annular dilatation or leaflet damage was not present and the patient had complete regression of symptoms, watchful clinical follow-up was warranted, and she was dismissed from the hospital on the eighth postoperative day.

Histologic examination with routine stainings and immunohistochemical (immunoperoxidase) techniques diagnosed a giant SFT. The huge epicardial tumor had a short (0.5 cm) and broad (4 – 2.8 cm) tumor stalk (Fig 1D) and was microscopically composed of a patternless and bland spindle cell proliferation with varying proportions of collagen bundles, rare mitoses (<2/10 high-power field), and low to intermediate cellularity (Fig 3A, B). No cell necrosis was found, whereas the inner core of the mass showed hyaline, acellular areas. No histologic evidence of neoplasia was found on the cauterized resection margin; few and scattered neoplastic cells were present ≥1 mm from it. Immunohistochemistry showed diffuse staining with CD34 (which is lacking in low-grade fibromyxoid sarcoma), vimentin, and Bcl-2 (Fig 3C, D), and negative S-100 protein (positive in schwannoma), ALK-1 (typical of inflammatory myofibroblastic tumor), HHF-35 smooth muscle cell actin (positive in fibromatosis and in myogenic and myofibroblastic tumors), and pancytokeratin AE1/AE3 (which is expressed in mesotheliomas). At 1-year follow-up, the patient is free of symptoms and signs of tumor recurrence, with two-dimensional echocardiography showing mild mitral valve regurgitation.

Comment
Solitary fibrous tumors have been described in both thoracic (pleura-based) and extrathoracic (liver, kidney,
prostate, central nervous system, periosteum, vagina, or skin) sites, the cardiac localization being very rare [1, 6–8]. Giant SFTs have been mainly diagnosed in pleura and in all ages and exceptionally in the heart [2, 5]. Most SFTs, in particular extrapleural tumors, except for those of mediastinal origin, are benign, although rare malignant variants have been reported so far, including a cardiac SFT [6, 8]. Recurrence is observed in 63% of all malignant SFTs and in 2% of benign tumors. The site, growth pattern (larger tumors are considered more likely to be malignant), and histologic features correctly identify the malignant cases, but a small subset behaves in an unpredictable fashion, the prognosis depending also on complete tumor resection [8]. A main task of histologic investigation is identifying tumors that have malignant potential; immunohistochemistry has a diagnostic role, CD34 positivity being necessary for diagnosis, whereas no specific molecular test is available.

In the present giant SFT, histologic evidence of malignancy was lacking, only a few areas of intermediate cellularity were shown by throughout examination, mitoses were very rare or absent, the tumor did not show any infiltrative feature, and clinical symptoms were related to the large size with displacement of the heart. The tumor was thoroughly excised macroscopically, and the tumor resection margin was microscopically free of
neoplasia, although a few scattered neoplastic cells were found quite close to it. We planned thorough follow-up at regular intervals; although the patient is well 12 months after the operation, this is not proof of cure, and we are aware that benign SFTs may recur [8]. Adjuvant chemotherapy was not undertaken, given the lack of evidence of tumor malignancy in the present case and the absence of an assessment of the role of adjuvant therapy in SFT; chemotherapy is usually considered in malignant sessile SFTs, particularly in recurrences, whereas it is not recommended in histologically benign tumors or in pedunculated malignant tumors [8].

In conclusion, giant SFT of the epicardium is exceptionally rare; histology and immunohistochemistry are mandatory for diagnosis and prognosis; because of the rarity of these tumors and of the unpredictable behavior of some SFTs, close follow-up is required, the management of such tumors representing a clinical challenge.

References


