REVIEW PAPER

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Cardiac solitary fibrous tumor – an extremely rare but potentially fatal diagnosis

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Abstract

The background of this review is a description of the case of a 28-year-old man with an extremely rare cardiac solitary fibrous tumor (SFT). Although this tumor was removed surgically and in the 6-month follow-up examination no relapse was noted, recurrence was observed and confirmed in the magnetic resonance imaging 4 months later. SFT prevalence, symptoms and signs, treatment options and prognosis are reviewed.

Key words: primary cardiac tumors, solitary fibrous tumor, surgery, follow-up, recurrence

Introduction

Primary cardiac neoplasms are relatively rare indications for surgical intervention, and among them the vast majority are benign, with the left atrial myxomas being most prevalent [1]. Outcomes following surgery for the latter are almost perfect and long-term survival in many of them is even similar to the general population [2]. Contrary to them, malignant cardiac tumors are much rarer but prognosis remains poor irrespective of therapeutic modality. A very special group of cardiac or pericardial neoplasms is the solitary fibrous tumors (SFT). They are extremely rare ubiquitous mesenchymal tumors composed of spindle or ovoid cells accompanied by variably collagenous stroma and hyalinized staghorn-shaped blood vessels [3–5]. They usually originate from the pleura, more often from the visceral than the parietal part [6, 7]. Cardiac localization of SFT is extremely uncommon.

In this paper, we would like to share our experience with malignant cardiac SFT treated surgically and collect current knowledge regarding symptoms and signs, diagnosis, therapeutic options and outcomes.

Case report

A 28-year-old man was admitted to the cardiac surgery department due to symptomatic pericardial effusion. On admission, the patient complained of dyspnea on exertion (classified as class II according to the NYHA functional classification) and general fatigue accompanied by fever for the last 2 months. Allergy to penicillin was reported. The patient denied smoking, alcohol addiction and drug usage. Physical examination on admission revealed only poorly accentuated muffled cardiac tones and moderate peripheral edema. Circulation was stable with the heart rate 60 beats per minute and blood pressure 120 over 70 mm Hg. The electrocardiogram (ECG) at rest was normal.

Computed tomography (CT) revealed massive fluid effusion in the pericardial cavity (up to 40 mm). Additionally, at the inferior cardiac wall, left lateral to the inferior vena cava a large nonhomogeneous mass measuring 62 × 13 mm in cross-section was found. Visible small capillaries inside the mass were also disclosed. Magnetic resonance imaging (MRI) confirmed a huge volume of fluid accumulated in the pericardial sac and a tumor adjacent to the inferior caval vein (Figure 1 A). The center of the pathological mass was enhanced after contrast agent administration. Within the tumor, there were zones with high and intermediate signal intensity in T2-weighted projections. Zones with very low signal intensity in phase sensitive inversion recovery (PSIR) after contrast were also detected. The lesion was connected with the intrapericardial part of the inferior vena cava penetrating through the pericardial sac. Further sections of the inferior vena cava were tumor-free. The tumor was richly vascularized.

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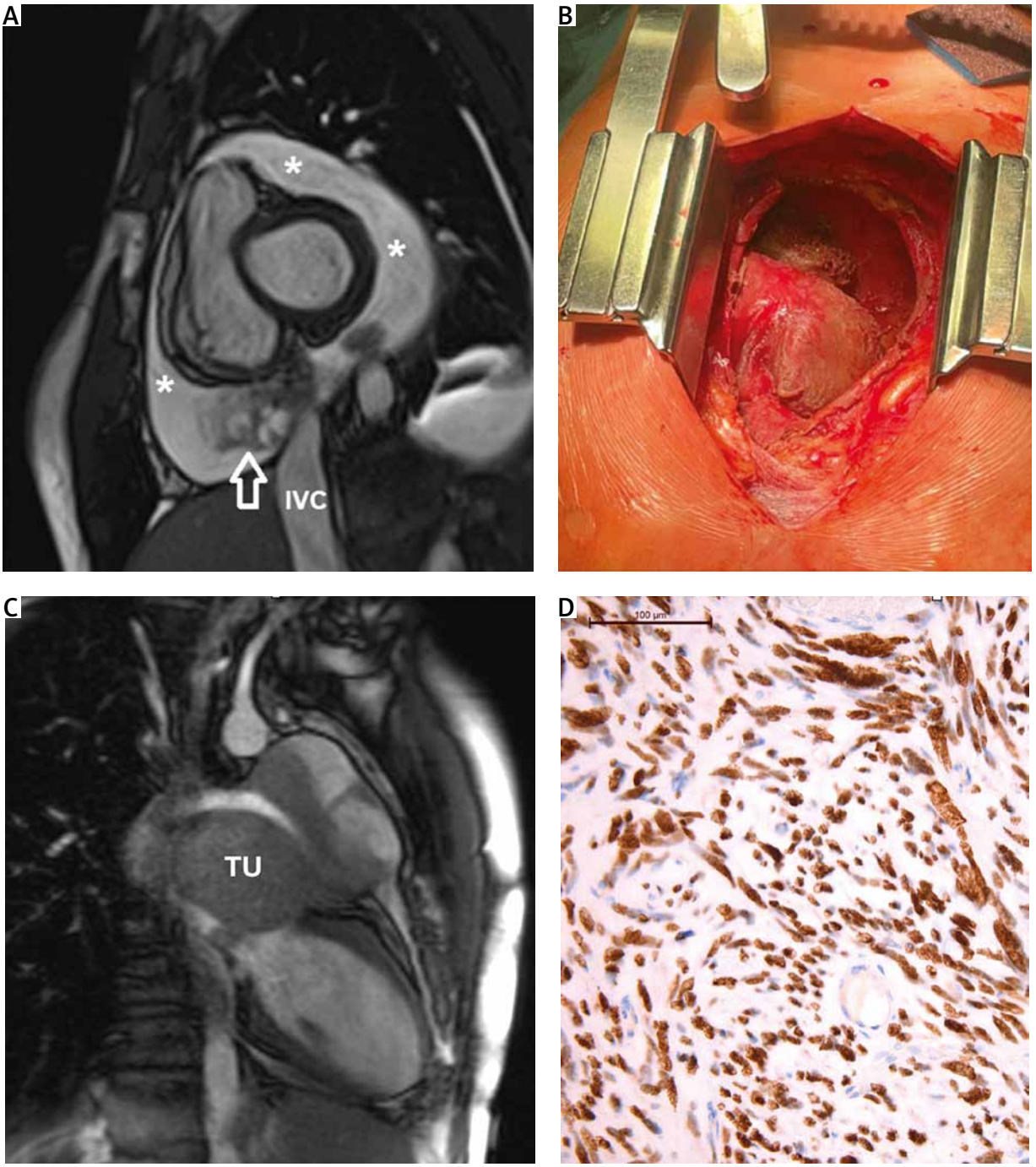

Figure 1. Cardiac solitary fibrous tumor. At the time of diagnosis, a solid tumor visualized by means of MRI (**A**) adhered to the inferior caval vein (IVC) and caused massive pericardial effusion (*). The tumor location was confirmed during surgery performed from full median sternotomy (**B**). Histopathological examination revealed a non-epithelial spindle cell tumor consisting of cells with various degrees of atypia, a high mitotic index and positive immunohistochemical staining for STAT6, vim, Bcl2, CKAE1/AE3 and ERG (**D**). Unfortunately, 10 months later recurrence of a tumor (TU) infiltrating the left atrium and tissues adjacent to the great vessels was visualized in MRI (**C**)

Surgery was performed from median sternotomy. The pericardium was thick and macroscopic signs of active inflammation were observed. Four hundred milliliters of brown fluid were drained from the pericardial sac. Fibrin was present on the surface of the heart. The tumor was located inside the pericardial sac and was attached at the place of the inferior caval vein and the right atrium entrance. After cardio-pulmonary bypass was implemented the heart was arrested with cardioplegic solution. The tumor was removed with the parts of both right atrial and inferior caval vein walls (Figure 1 B). Further intraoperative steps were standard. Pericardial effusion and tumor specimens harvested intraoperatively were sent to the histopathological laboratory. The tumor was a non-epithelial, spindle cell neoplasm with cells organized in various systems (Figure 1 D). The herringbone pattern was dominant,

although bundle one was also partially present. Microscopic examination showed moderate cellular atypia. The mitotic count was 9/10 high-power fields. Necrosis constituted 15% of the tumor. Immunohistochemical staining revealed that its cells expressed positively for signal transducer and activator of transcription 6 (STAT6), vimentin (vim) and Bcl2. The final diagnosis of SFT was established and following the uneventful postoperative period the patient was referred to outpatient (both oncological and surgical) clinics. The oncologist on the basis of the findings of all examinations decided not to administer adjuvant therapy.

At the 6-month follow-up visit, the patient remained asymptomatic with echocardiography without any signs of tumor recurrence. Unfortunately, 4 months later dyspnea on exertion occurred and imaging studies again visualized a huge tumor originating from both the peri- and myocardium. This time, the signs of right ventricular hypertrophy in ECG were noted. In the echocardiographic examination a pathological mass markedly impairing blood flow was noted. Massive infiltration of the left atrium and tissues adjacent to the ascending aorta and pulmonary trunk but not distal metastases were confirmed in MRI (Figure 1 C). The heart team decided to postpone surgery and referred the patient emergently to the chemotherapy department.

Solitary fibrous tumor (SFT)

Prevalence, anatomy, biology

SFT is an uncommon soft tissue derived neoplasm originating from mesenchymal tissue of unknown etiology [5, 6]. It usually arises from the pleura or abdominal cavity, and less frequently from the mediastinum, extracranial tissue, pelvis, or retroperitoneum [6, 8]. In our case it originated from the inferior caval vein to the right atrium outlet, which must be considered as exceptional [9].

SFTs may be present in both genders in adult persons regardless of their age, yet they are most commonly diagnosed in patients in their 5^{th} or 6^{th} decade of life [3–5]. The youngest reported subject diagnosed with cardiac manifestation of SFT was a 5-month-old male infant whereas the oldest one was an 81-year-old man [6]. According to our knowledge there were only 2 cases with cardiac SFT younger than ours [10, 11]. Unfortunately, young age seems to be a negative predictor of SFT survival. Although surgery to excise the tumors was successful in both cases, the patients died a few months later due to either pneumonia or uncontrolled tumor dissemination [10, 11]. In our case recurrence 10 months following primary surgery was observed.

STAT6, Bcl2, CD34, CD99 and vim immunohistochemical staining is commonly used for definite verification of SFT [3]. Immunoexpression of STAT6 on tumor cells plays a key role in SFT pathological diagnosis and features both high sensitivity and specificity [3–6, 12, 13]. It can also be used as a useful tool to differentiate SFT from its histologic imitators [13]. Bcl2, vim, CD34 and CD99 are also frequently expressed on tumor cells but they are less specific [3, 6, 8]. In our case STAT6, Bcl2 and vim staining were positive.

Symptoms and signs

SFTs frequently manifest no symptoms and cause no pain, grow gradually and are usually benign [4, 14]. Thoracic SFTs are often found incidentally during chest imaging [3]. Patients with cardiac SFTs present with a varying range of complaints including exertional and resting dyspnea or less commonly fatigue, chest discomfort, dry cough, heart palpitations and generalized edema. Sometimes they are asymptomatic. The type of symptoms and their presence depend on the localization and the size of the tumor [6, 9, 15]. The patient described herein experienced breathlessness on exertion, chronic fatigue and fever.

Treatment options

Although there are no established guidelines for SFT treatment, surgical excision of the tumor is indicated as a method of choice [3, 7, 8, 16]. It is vital to obtain adequate negative margins as it has decreased the rates of metastases and local recurrence [3, 16]. In most cases this kind of treatment may provide the patient with a favorable outcome [16]. In tumors with a histologically malignant component, surgical excision may not be sufficient for the cure [16, 17]. Additional treatment might include radiotherapy and chemotherapy [3, 8]. Antiangiogenic drugs (pazopanib is recommended in the first line) improved outcomes in SFT patients. They were also shown to have superior efficacy over other cytotoxic drugs [8, 18]. In our case surgical treatment was implemented without any adjuvant therapy. Long-term, even lifetime systematic assessment is necessary because cases of disease recurrence after a long period of time were reported [3]. Due to the nature of the tumor and its rarity, a multidisciplinary team should take care of these patients. Quite often an individualized decision must be taken because there is no commonly accepted therapeutic management for SFT [3, 8].

Prognosis

In most cases cardiac SFTs are benign, yet sometimes malignancy is present. Out of 22 reported cases since 1987, 14 (64%) have been benign SFTs and 5 (23%) have featured malignancy, whereas for the remaining cases the information is not available [6]. The association between histology and clinical course of the disease is highly unpredictable [5]. The latest WHO classification regarding SFT recommends risk stratification models over the traditional benign/malignant distinction, which was based on microscopic findings [5, 8]. The most commonly used model for metastasis risk and mortality comprises patient age, mitotic count and necrosis percentage in the tumor (necrosis predicts only the metastasis risk, not the mortality risk) [4, 19]. Small tumors with low mitotic rates are not predicted to cause patient death and metastasize while tumors in 55-year-old people and older, bigger than 15 cm in diameter and with 4 or more mitotic figures/10 high-power fields have increased mortality and elevated risk for metastatic disease [19]. This viewpoint enables the perception of SFT as a bio-

logical continuum rather than as a neoplasm divided into separate types [18]. The number of mitotic figures in our case was high but the patient was young and the mass was not large. Therefore our patient was considered to have intermediate risk for development of metastases [5]. The general relapse rate in SFT patients is 10 to 30%, with higher prevalence within the first 5-year period and rare recurrences reported within the next 10 years of follow-up [4]. Five-year probability of survival varies between 59 and 100%, whereas 10-year survival varies between 73 and 89% [3].

Conclusions

Cardiac SFT is an extremely rare but challenging disease in terms of diagnosis and optimal management. Although it is most commonly found in middle-aged patients, younger people may also be affected. STAT6 is the best marker for SFT histopathological diagnosis. Surgical excision with adequate margins is the recommended treatment mode. Unfortunately, the prognosis seems to be unpredictable.

Disclosure

The authors report no conflict of interest.

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