Case report - Cardiac general

Right atrial solitary fibrous tumor – a new cardiac neoplasm?

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Abstract

Heart tumors are very rare neoplasms. Solitary fibrous tumors are mesenchymal neoplasms first described in 1931 as pleura-based lesions. We are the first to describe the clinical symptoms, surgical technique for removal, histology, and exact location as well as medium-term follow-up of a solitary fibrous tumor in the heart.

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1. Introduction

Heart tumors are very rare neoplasms. Solitary fibrous tumors (SFT) are mesenchymal neoplasms first described in 1931 as pleura-based lesions [1]. However, in recent years extrapleural solitary fibrous tumors have been increasingly detected [2]. Although most SFTs are benign, they can recur and malignant variants are reported [3]. We are the first to describe the clinical symptoms, surgical technique for removal, histology, and exact location as well as medium-term follow-up of an SFT in the heart.

2. Clinical presentation

A 39-year-old woman with a history of palpitations for 3–4 months was admitted to the hospital. She had been previously healthy. Results of physical examination were within normal limits. Holter monitoring showed a normal sinus rhythm with multiple supraventricular extrasystoles. Echocardiography demonstrated a 4.4 × 4.5 × 5.5 cm hyperdense, polypoid structure with a broad adherence to the free wall of the right atrium (Fig. 1A). Based on the echocardiographic presentation, the diagnosis of an intracardial tumor was made which was removed during heart surgery with cardioplegic arrest. Fig. 1B shows the tumor in situ. The tumor was located at the junction of the free atrial wall and the cranial part of the atrial septum. During the procedure, all visible parts of the tumor were excised. Fig. 1C shows the excised specimen. The created defect was closed with autologous pericardium.

Twelve months after surgery the patient was free of symptoms. Holter monitoring showed a normal sinus rhythm without supraventricular extrasystoles. Echocardiogram twelve months after surgery revealed no pathological findings.

3. Pathological findings

Macroscopically, the tumor had a maximum size of 5.6 cm in diameter and showed small fibers of myocardium along the edge of the resection area. On the cut surface the mass was white with slightly curling fibers of firm consistency. No major hemorrhage or necrotic features were present.

Histologically, monomorphic spindle cells intermingled with broad collagenous fibers and a slightly alcinian blue positive myxoid matrix. No mitotic figures were seen. Some spindle cells were growing focally between the cardiomyocytes present on several resection areas (Fig. 2A–D).

Immunohistochemically, the tumor cells stained strongly cytoplasmically for CD 34 and vimentin, to a lesser extent for bcl-2 and only rarely for alpha-actin (Fig. 2E–F) and CD...
Fig. 2. Histology of the SFT showing spindle cells within a fibre-rich and myxoid matrix. A, B: H&E stainings. Asterisk indicates right atrial myocardium with ingrowth of tumor cells. C: EvG staining highlighting the collagen content. D: Alcian-blue staining showing the light myxoid matrix. E: Strong cytoplasmic staining for CD 34 and F: bcl-2, lesser intense cytoplasmic staining of G: Vimentin and H: α-actin.

99. Other markers like CD 68, desmin, cytokeratine, S-100, neuron specific enolase (NSE) gave negative results (data not shown).

4. Discussion

Odin et al. [4] claim that Columbus of Padua was the first to describe a cardiac tumor in 1559. Clarence Crafoord [5] was the first to excise a cardiac tumor on cardiopulmonary support in 1954. However, tumors of the heart are uncommon. Autopsy series report an incidence of between 0.001–0.03% in the general population [6].

Most cardiac tumors are benign, accounting for 70–80% of all tumors that originate in the heart. Most of them are myxomas. Fibromas are seldom reported.

Solitary fibrous tumor, also referred to as fibrous mesothelioma, submesothelial fibroma and localized fibrous tumor, was first described in 1931 [1]. Initially, SFT was described as a pleura-based neoplasm and the diagnosis was based on histomorphological characteristics.

The microscopic picture of an SFT has two common patterns. The first is described as a pattern of bland, wavy-appearing spindle cells in a collagenized stroma. Some of the SFTs may be hypocellular or hypercellular, or both. The second common pattern is ‘hemangiopericytoma-like’, where the lesional cells are densest around branching blood vessels. Because of this variable histological structure SFTs might be mistaken for other tumors such as hemangiomyomas or fibromas. However, recent immunohistochemical tests (CD-34, bcl-2 oncoprotein, CD-99 [7–9]) allow to separate SFTs from tumors with similar histomorphological characteristics. This may be the reason for the increasing number of SFTs diagnosed, that have been reported in liver, kidney, prostate, central nervous system, peristomeum, vagina or skin [2] recently. SFTs have also been reported in the heart in either pericardial or epicardial locations [10]. The UCLA group [4] categorized 30 cardiac tumors, only one of which was an SFT. However, no information on its exact location or histology was provided. We are therefore the first to describe an SFT in the right atrium.

Most SFTs are benign. Atypical and malignant variants are rare and have been reported in thoracic and extrathoracic locations. The atypical and malignant variants tend to recur and metastasize. The histologic features of malignancy include increased mitotic activity, nuclear pleomorphism, necrosis and, as the only feature of malignancy that was found in our case, high cellularity. A malignancy of our tumor seems therefore unlikely.

Complete local excision of SFTs is curative in most cases. However, a wide local excision of the tumor can be problematic in the heart. In our case, the SFT was thoroughly excised microscopically, but tumor cells were observed growing microfocally between the atrial cardiomyocytes. Tumor recurrence is therefore possible. Despite an uneventful postoperative course, we consequently performed a thorough follow-up of this patient 12 months after surgery, and we will continue to examine her at regular intervals.

In conclusion, this is the first report of an SFT as a right atrial neoplasm. However, the diagnosis of previous SFTs may have been missed due to a lack of accurate immunohistochemical tests. SFT is therefore certainly a rare, but not necessarily a new cardiac neoplasm.

References