



Case Report

Benign cardiac schwannoma: A case report

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ABSTRACT

We present an interesting case of a 47-year-old female with an incidental finding of a cardiac tumor located between the right atrium and right ventricle. After some time, the patient became symptomatic, the mass was excised, and pathological studies revealed it to be a benign schwannoma. Schwannomas are tumors of nerve sheaths commonly found in cranial and peripheral nerves. Their occurrence in the heart is exceedingly rare.

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1. Case report

The patient was a 47 year old woman with a history of ovarian cancer that was treated previously. During the work-up for her ovarian cancer, a large pericardial mass was discovered. Because she was asymptomatic at the time, close monitoring of the growth of the tumor was suggested for the time being.

Subsequent CT scans (Fig. 1), described the lesion as a lobulated soft tissue mass measuring $9.6 \times 7.1 \times 7.6$ cm that was located at the inferior surface of the heart. An echocardiogram indicated that there was some compression of the right atrium and the right ventricle, and areas of calcifications and echolucent areas suggestive of cystic features were found.

She began noticing symptoms including sharp chest pain, occasional shortness of breath on incline or stairs (NYHA I or II) as well as 2–3 episodes of palpitations per month lasting 2–3 min. Due to these symptoms she elected to have surgery to remove the tumor. Intraoperatively it was discovered that the blood supply of the tumor came from a branch of the right coronary artery (RCA) and the posterior descending artery (PDA) which coursed through the tumor. The operation was carried out successfully and consisted of a resection of the tumor, ligation of the PDA and a saphenous vein graft for a coronary bypass to the distal PDA.

Grossly, the tumor was an ovoid mass measuring $9.0 \times 7.0 \times 6.5$ cm, with a fresh weight of 160 g. The outer surface was grey-tan and smooth with a lobulated appearance. The cut surface was multi-lobulated with areas of pale tan, firm tissue and sanguineous fluid (Fig. 2).

Microscopically, the representative sections show a thick, fibrous capsule surrounding the tumor. A biphasic pattern of compact spindle cells (Antoni A) and loosely formed hyper cellular areas (Antoni B) with thick walled hyalinized vessels were appreciated. Verocay bodies, formed by palisading cells were also occasionally identified. There was no evidence of necrosis. There were no cellular pleomorphism and no atypical mitoses identified (Fig. 3).

Immunohistochemistry was performed and the cells are strongly positive for S100, and negative for desmin, calretinin, caldesmon, and alpha smooth muscle actin (Fig. 3). The cells that line the vessels in the tumor are positive for CD31 and CD34. Based on the gross and microscopic evidence a diagnosis of benign schwannoma of the heart was made.

2. Discussion

Cardiac tumors are, in general, rare. The estimated frequency of cardiac tumors ranges from 0.0017 to 0.33%. Three-quarters of these tumors are benign and nearly half of the benign tumors are myxomas [4]. Only 17 cases of primary cardiac schwannoma were reported previously in the literature and the majority of these cases were malignant. It is believed to originate from the cardiac plexus, or the cardiac branch of the vagus nerve. It is located primarily on the right side of the heart, particularly in the right atrium. However, schwannomas can also be found

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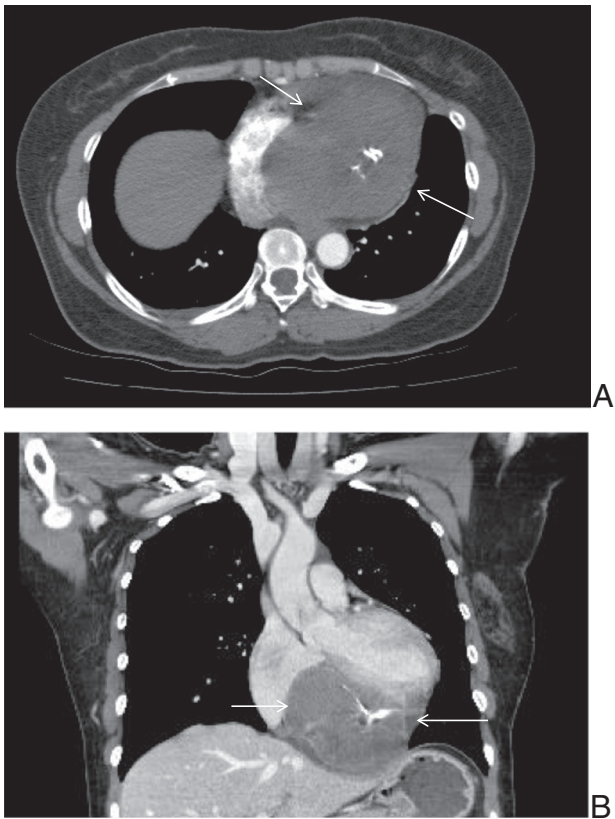


Fig. 1. Axial (A) and coronal (B) contrast-enhanced CT through the thorax identifies a well-defined, lobulated mass (indicated by arrows) with central coarse calcifications and faint enhancement. The lesion exhibits mass effect on the adjacent cardiac chambers with inversion of the diaphragm. No evidence of localized invasion or pericardial fluid.

in the left atrium, right ventricular outflow tract, and/or atrioventricular groove [5,7].

In asymptomatic patients, as in our case, diagnosis is usually incidental as a result of an imaging test obtained for an unrelated reason. In patients with symptoms, clinical presentation depends on the size of the mass, anatomic location, and friability or ability to embolize. Most

commonly patients present with cardiovascular symptoms including chest pain, fainting, cardiac failure, valvular disease-like symptoms and pericarditis symptoms, retention of pericardial effusion, cardiac tamponade, arrhythmia, and conductive disorders. In our case, the patient was asymptomatic during the initial tumor discovery. She subsequently developed symptoms with the progression of the disease and enlargement of the tumor [6].

Cardiac masses are primarily visualized and monitored by various imaging techniques. Echocardiography is usually the first modality used due to its non-invasive nature. It can quantify the tumor size, shape, attachment, and mobility, and can screen other cardiac chambers accurately for additional tumors. However, MRI offers a high spatial resolution and is the best modality for characterizing tumor tissue and vascularity. When MRI is not indicated, for example in patients with claustrophobia or carrying a pacemaker or defibrillator, then computed tomography (CT) is an alternative. CT can be used to locate and identify calcifications and to visualize the mass in relation to the coronary arteries. It is not possible to differentiate between different types of cardiac tumors on imaging [2].

A definite diagnosis of cardiac tumors can only be completed after histological examination of samples taken at autopsy or surgical resection. Histological features of a schwannoma include the biphasic architecture of Antoni A and B patterns, as well as nuclear palisading (Verocay bodies) and fibrous capsules containing the cells derived from the nerves. The Antoni A pattern contains elongated fascicles in the areas of moderate-to-high cellularity with a small stromal matrix. In the Antoni B pattern, the tumor is less densely cellular with a loose meshwork of cells along with microcysts and myxoid changes. It is considered benign based on the absence of cytological atypia, mitotic figures, and necrosis [3].

Immunohistochemistry can complement histologic findings. The S-100 protein is a specific marker for schwannoma. It is obtained from the neural crest origin tumors from which the melanocytes and Schwann cells are derived [1].

The prognosis of benign cardiac tumor depends on resectability. After complete resection the prognosis is excellent, and adjuvant therapy is not needed [4]. If surgery is not indicated, chemotherapy can be an alternative. In our patient, chemotherapy given for ovarian cancer did not affect the size of the cardiac schwannoma that continued to grow, and the patient became symptomatic thus surgery was performed. Hereby we present a case report of a rare benign primary schwannoma of the heart.



Fig. 2. The cut surface of the cardiac tumor showing multi-loculation, with areas of pale tan, firm tissue, and cystic spaces with sanguineous fluid.

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