Case report

Vascular complications associated with a large cardiac fibroma

Francesco Patane*, Edoardo Zingarelli, Alessandro Verzini, Michele di Summa

Cardiac Surgery Department, 'San Giovanni Battista' Hospital, Turin, Italy

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Abstract

A case is reported of a 30-year-old patient with an intrapericardial tumour with heart failure. After the diagnostic protocol, surgery was performed initially without extra-corporeal circulation (ECC). Due to the location, size and to the large connection with the most important vascular structures, the ascending aorta ruptured accidentally during resection and was replaced after using ECC in emergency. The post-surgical course was regular and the tumour was identified histologically as a fibroma. © 2001 Elsevier Science B.V. All rights reserved.

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1. Introduction

Primary cardiac tumours are rare. Their incidence ranges from 0.0017% to 0.28% in autopic series. Among malignant tumours, 10% of primary cardiac tumours, sarcomas are frequent. Among benign cardiac tumours, the most frequently occurring are myxomas [1]. Fibromas are the second most common tumours in infancy in autopic series, after rhabdomyoma [2]. Normally, they do not invade, and they can become large, as in the case reported. This characteristic must be contemplated to decide on a correct surgical approach. In large tumours the most important surgical difficulties consist in finding a cleavage plane and following it without damaging vascular and cardiac structures, and trying to resect the mass whole. The electro-corporeal circulation (ECC) is an important help to obtain these aims.

2. Case report

A 30-year-old man was hospitalized in September 1999 at our Department, presenting epigastric distension with facial edema on awakening, edema of the legs on standing, and other signs of heart failure. Objective examination revealed increased S2P and presence of a holosystolic murmur of 3/6 intensity audible in all areas.

The electrocardiogram showed low voltages in the precordial and limb leads, and subsequently revealed a high takeoff of the ST segment from V1 to V3 with inverted T.

Chest radiography revealed a clearly-outlined mass in the left hemithorax which was indisociable from the mediastinic and cardiac profiles.

Transthoracic and transeosophagus echocardiography showed: an enlarged right atrium; a hypertrophic right ventricle with continent tricuspid; a thickened pericardium; on the cardiac base, an echorelecting formation, probably intrapericardial, compressing the pulmonary trunk and stenosing the right ventricular outflow with systolic gradient 55–58 mmHg.

The thoracic-abdominal CAT scan with contrast medium showed pericardial effusion 80 mm thick, close to the cardiac base, extending to the aortic arch with compression of the left pulmonary parenchyme.

Cardiac NMR (Fig. 1) before and after gadolinium administration showed a solid formation with distinct margins in the anterior mediastinum, probably extrapericardial, 8.8 × 7.7 × 7 cm. In T1 the formation appeared isointense; in T2 it appeared non-uniform with little enhancement after gadolinium administration. The mass extended from the emergence of the innominate artery and of the left subclavus to the anterosuperior mediastinum, reducing the right ventricular outflow and surrounded three sides of the ascending aorta without altering the lumen. The dislocated pulmonary trunk was without stenoses. The mass compressed the right ventricle outflow and the pulmonary artery trunk.

The tumour was removed surgically.

With an open pericardium, the formation appeared as a solid, intrapericardial, yellowish–white oval mass about
10 × 10 cm. After having examined the mass and its relation to the surrounding structures, the peduncle appeared to originate from the aortapulmonary window, and there appeared to be a favourable cleavage plane for its removal without the help of the ECC. Having reached the area of the aortapulmonary window, during dissection of the mass from the tunica adventitia of the aorta, sudden laceration of the ascending aorta required emergency femoro–atrial ECC. Having completed tumour resection, clamped the ascending aorta and performed cardioplegia, the ascending aorta was replaced with a prosthetic tube.

Macroscopic examination showed a firm elastic mass weighing 450 g, capsulated, 10 × 7 × 7 cm, maximum diameter 12 cm (Fig. 2). On dissection it appeared fasciculated and free from areas of necrosis.

Microscopic examination, performed in our Pathological Center, showed spindle cells in proliferation, mixoid areas and poorly vascularized collagenized areas. The immunohistochemical reactions were positive to antivimentine antibodies and negative to anti-S 100 and anti-smooth and striated muscle actin antibodies. Among the spindle cells there were accumulations of histiocytes positive to anti-CD 68 antibodies. These findings were compatible with the diagnosis of fibroma.

After a normal postsurgical course, the patient left intensive care on day 2 and was dehospitalized on day 7. Echocardiography immediately after surgery and at 6 months was normal.

3. Discussion

In the past, cardiac tumours were most frequently observed upon autopsy, though today, thanks to developments in the diagnostic and therapeutic fields, they are often better managed.
Fibromas do not metastasize, do not infiltrate, but they cause ventricular tachyarrhythmia, heart failure, sudden death; anginal pain is rare at presentation. The ratio between pediatric patients and adults is 3:1, and about 75% are diagnosed by age 2. Heredity and gender are not relevant [3,4]. The congenital origin of the tumor is probable and it is frequently identified without symptoms.

Electrocardiography reveals anomalies of the ST tract, the T wave, signs of ventricular hypertrophy, conduction defects, deviation of the cardiac axis, and abnormal Q waves [3,5,6].

Thoracic radiography reveals alterations of the cardiac profile.

Echocardiography provides data on the location, size and cardiac functionality, and is also useful for prenatal diagnosis [2].

Thoracic CAT scanning and MRI are necessary for presurgical evaluation, while angiography determines filling defects, alterations in shape and associated pathologies.

In the case reported the cleavage plane, salvaged by opening the pericardium, which was in the aorta-pulmonary window, led us to avoid the ECC. Nevertheless when the tumor is large and the connections with vascular structures are important the ECC is an effective remedy because it allow us to greatly reduce surgical risks and at the same time to have a radical resection of the mass. This is also true in those cases where the lack of infiltration could lead to avoid ECC.

In the case reported, the ascending aorta was found to be very thin even if there was no infiltration. During the dissection of the mass from the aorta-pulmonary window a slight tearing of the aortic adventitia occurred. Then the tear extended because of the traction of the mass. So we had to use the ECC in emergency.

We think that the use of the ECC is especially essential in the resection of malignant tumors, because in these cases the infiltration of vascular structures of the mediastinum is frequent but also in the resection of giant benign tumors which, even if they don’t invade, damage surrounding structures around because of their bulk.

References