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Papillary fibroelastoma of the left atrium in a 3-year-old boy

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Abstract

A 3 year old boy presented with a cardiac murmur. Except for mildly impaired physical condition that was more likely due to asthmatic bronchitis, he was asymptomatic. Cardiac echocardiography disclosed a large tumor in the left atrium originating from the interatrial septum. The patient was immediately referred to surgery for excision of the tumor. Histological examination of the tumor showed a typical papillary fibroelastoma. The occurrence of a papillary fibroelastoma in a child is a rarity.

MeSH: cardiac papillary fibroelastoma, infant

Introduction

Primary tumors of the heart in infants and children are rare. Among pediatric admissions cardiac tumors have an incidence of only 0.0017% - 0.08%.[1] In children the most common primary cardiac tumors are rhabdomyomas (45 %) and fibromas (25 %), followed by myxomas, teratomas, and haemangiomas.[2] Cardiac papillary fibroelastomas (CPFs) are

extremely rare in children. In adults they represent after myxomas (30%) and lipomas (10%) the third most common benign primary cardiac tumor (7.9%).[3] Approximately 90% of the CPFs are attached to valves.[3,4] Most CPFs do not cause symptoms and are usually incidental findings by routine echocardiography or at autopsy. However, early diagnosis of this condition is important, since it represents a surgical correctable cause of systemic emboli, stroke, myocardial infarction, and sudden cardiac death.[4–11] We report the case of a 3 year old boy who presented with a cardiac murmur, caused by a tumor in the left atrium originating from the interatrial septum.

Case report

A 3 year old boy presented at our heart centre with a cardiac murmur that had been noticed first 2 weeks prior to admission. The mother described a mildly limited physical activity most likely due to asthmatic bronchitis. There was no history of chest pain, oedema, cyanosis, or loss of consciousness. On physical examination, he presented in a good general condition. His pulse rate was 150 bpm and regular, and the blood pressure was 75/50 mmHg. A 2/6 mid-systolic murmur was audible above the heart apex. No definite splitting of the second heart sound was detected. The lungs were clear on auscultation with good bilateral air entry. No hepatosplenomegaly. There was no peripheral oedema. His haemoglobin was 6.9 mmol/l, the haematocrit 33 %. White blood cell counts and differentiation, platelet count, creatinine, ASAT, C-reactive protein, coagulation parameters were within normal limits. The electrocardiogram was unrevealing. Chest X-ray films showed a slightly increased size of the heart silhouette and moderately increased pulmonary vascular markings.

Transthoracic echocardiography disclosed a tumor in the left atrium that arose with a stalk from the upper part of the interatrial septum reaching a size of 3×3×2 cm. In diastole the tumor prolapsed through the mitral valve into the left ventricle (Figures (Figures11 and and22).

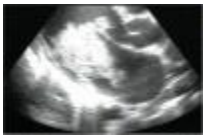

Figure 1
Transthoracic echocardiography, parasternal long axis.

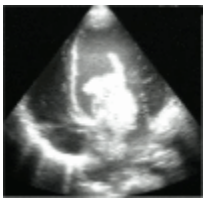

Figure 2
Transthoracic echocardiography, four chamber view.

The tumor had a flaccid structure and slipped partially with the blood flow back and forth through the mitral valve into the left ventricular inflow and outflow tract. The moving images convey the impression of potential embolization.

The patient was immediately referred to surgery for excision of the tumor. The operation was performed under cardiopulmonary bypass. The left atrium was opened. There was a 3×3×2 cm colloid mass originating from the atrial septum. The tumor had macroscopically the shape of a sea anemone. Histopathological examination revealed a typical papillary fibroelastoma. The tumor was fully resected including its attachment, and the defect in the atrial septum was closed by a pericardial patch. The postoperative course was uneventful.

Discussion

Papillary fibroelastomas represent 7.9% of benign primary cardiac tumors in adults.[4] More than 90% of CPFs are solitary;[3,4] CPFs present rarely as multiple lesions.[12,13] Approximately 90% arise from valvular tissue, most commonly from aortic or mitral valves.[3,14] There are only few reports in the literature of papillary fibroelastomas in young children. In an early report on papillary fibroelastomas was only one patient below the age of twenty years.[3] De Menezes et al. reported a case of a 3.5-year-old boy with a stroke secondary to embolization of a mitral valve CPF.[15] A 10-year-old child presented with biventricular failure after an acute myocardial infarction due to a large CPF arising from the aortic valve.[8] In a study including 162 patients with CPF the age ranged from 5 to 86 (mean, 60± 16 years).[16]

On gross anatomical examination, CPF resembles a sea anemone, consisting of multiple fingerlike fronds radiating from a stalk. Microscopically, each frond consists of a collagenous and elastic core, surrounded by a mucopolysaccharide matrix, and covered by endothelial cells.[16,17]

The pathogenesis of CPFs remains under discussion, but several possible explanations have been reported, including previous mechanical damage to the endothelium, iatrogenic factors, organizing thrombi, and a latent infectious mechanism due to cytomegalovirus.[3,17–22] The occurrence in neonates and infants, as in the presented case, supports the hypotheses of hamartous origin or congenital abnormalities.[3,23]

With the advent of echocardiography, an increasing number of CPFs have been diagnosed during life. Typical echocardiographic features include the following:

1. The tumor is round, oval, or irregular in appearance, with well-demarcated borders and a homogeneous texture
2. Nearly half of CPFs have small mobile stalk/s.[16,24]

Sun et al. found that 99% of the CPFs were <20 mm in the largest dimension.[16] The largest reported CPF is 53 mm.[25] The CPF of the here presented boy showed echocardiographic features consistent with published case descriptions. However, the size of 30 mm is unusual.

Embolization of CPF fragments or an attached thrombus may cause cardiac, pulmonary, or neurologic symptoms, and prolapse of the tumor into a coronary ostium may cause sudden death.[4–11] Accordingly, prophylactic tumor excision with valve replacement when necessary, even in asymptomatic patients, is considered by many to be the treatment of choice.[4,10,15] Sun et al. recommend the following management of patients with CPF. Surgical removal of right-sided CPF in asymptomatic patients is indicated only for large mobile tumors. The presence of a patent foramen ovale with a sizeable right-to-left shunt is an additional consideration for right-sided CPF. Asymptomatic patients with small, left-sided, nonmobile (no stalk) CPFs are usually observed. However, CPFs ≥ 1 cm, especially, if mobile, should be considered for excision, including patients with other cardiovascular disease or young patients with low risk of surgery and a high cumulative risk for embolization.[16] Recurrence of a papillary fibroelastoma has not been reported.

We describe the case of a patient with CPF that was uncommon for the following reasons:

1. The patient was only 3 years old
2. The CPF was not arising from valvular tissue, but from the interatrial septum
3. The tumor exceeded the usual size of ≤ 20 mm.

Due to a cardiac murmur echocardiography was initiated and disclosed the tumor. Fortunately, excision was performed before a symptomatic embolization occurred.

Papillary fibroelastomas are extremely rare in children. Although symptoms related to CPFs are uncommon, there is considerable evidence that this tumor can cause life-threatening complications. Therefore, the diagnosis of a CPF in infants and children mandates prompt surgical resection of the tumor due to their high cumulative risk for embolization.

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