Intracardiac varix of the left atrium

Kaori Katoa†, Tetsu Yamaneb, Shoji Suzukic and Masahiko Matsumotoa

a Second Department of Surgery, University of Yamanashi Hospital, 1110 Shimokato, Chuo, Yamanashi 409-3898, Japan
b Department of Pathology, University of Yamanashi, Yamanashi, Japan

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Abstract

A 68-year-old female patient with a history of hyperlipidaemia and fatty liver was referred for evaluation of an incidentally detected asymptomatic cardiac mass. Computed tomographic scan imaging showed a large calcified mass in the left atrium. Echocardiography revealed a 2.4 × 1.5 cm, well-circumscribed, round, high echoic mass with severe calcification and low mobility attached to the lower rim of the fossa ovalis. The cardiac mass along with part of the fossa ovalis and left atrial wall were excised. Histological diagnosis was compatible with intracardiac varix.

Keywords: Cardiac varix • Atrium • Calcification

INTRODUCTION

Intracardiac varix is an uncommon tumour-like lesion of the heart, with the majority found in the right atrium at the lower rim of the fossa ovalis. Here, we report a case of successful surgical resection of a mass found in the left atrium that was diagnosed as an intracardiac varix.

CASE REPORT

A 68-year-old woman with a history of hyperlipidaemia and fatty liver was referred for evaluation of an asymptomatic cardiac mass incidentally detected by a computed tomographic (CT) scan of the chest. On admission, physical and neurological examinations were normal, and chest roentgenogram and electrocardiogram were within normal limits. Non-contrast cardiac CT image showed a large calcified mass in the left atrium with no extracardiac extensions (Fig. 1). Transthoracic echocardiography revealed a 2.4 × 1.5 cm, well-circumscribed, round, high echoic mass with severe calcification firmly attached to the lower rim of the fossa ovalis in the left atrium. No haemodynamic changes were observed, and serologic studies showed no evidence of coagulation abnormalities.

Resection was planned due to a large intracardiac mass. Under cardiopulmonary bypass, we approached the mass through a right atriotomy trans-septal incision. The mass had a pedunculated base at the lower rim of the fossa ovalis and was resected in one piece along with a part of the fossa ovalis and left atrial wall. The excised specimen was elastic firm, enveloped with a smooth surface, appeared dark bluish inside, and measured 2.3 × 1.4 cm (Fig. 2a). Upon sectioning the mass, round deposits of calcification surrounded by blood clots and connective tissue could be observed. Postoperative course was uneventful, and 1 year after operation, the patient is asymptomatic, and no signs of recurrence are found under echocardiography.

Pathologic studies revealed the mass to be a unilocular cyst lined by remnants of elastic fibre, filled with peripheral fibrin clots and focal calcium deposits, suggestive of phlebolith formation (Fig. 2b). These findings were consistent with cardiac varix.

DISCUSSION

Intracardiac varices are tumour-like lesions of the heart, with its incidence estimated to be ~0.07% from postmortem findings [1]. However, even more infrequent are the number of case reports. In fact, only nine cases in living patients have been reported up to date [2–10]. The low recognition of the disease and the fact they rarely grow large enough to develop symptoms may account for the difference between its incidence in autopsied heart and case reports.

Differential diagnosis is quite challenging. Pre-operatively, they are frequently misdiagnosed as myxomas. However, there are a few characteristics of varices that may allow us to distinguish this entity from other cardiac masses. First, varices are often accompanied by calcification or form cysts. Including our experience, five case reports showed calcification, three showed cystic changes, and two showed both cystic change and calcification. Incidence of myxomas accompanying calcification is far less common, and even more, are rarely described as ‘cystic’. Under echocardiography, myxomas are usually characterized to be mobile, homogenously solid masses, where varices usually have a fairly broad pedunculated base. In case of myxomas, the use of contrast-enhanced CT often reveals a well-defined spherical or ovoid mass with lobular contours, whereas a smooth surface seems to be a characteristic feature of varices. Furthermore, other masses such as haemangiomas usually show strong enhancement on contrast CT or show vascular blush on
angiography, and thrombi are usually accompanied by atrial fibrillation or coagulation abnormalities and occur in the atrial appendage. This suggests that a close examination of the echocardiographic and CT features and risk factors may enable us to suspect cardiac varices pre-operatively.

Pathologically, intracardiac varices are known to be endocardial, unilocular, blood-filled cysts lined by endothelial cells and organizing thrombi. Histological analysis may misdiagnose a varix to be a thrombus if endothelial cells are absent. One must take into account that, even without endothelial cell lining, thrombosed varices require close examination of remnants of elastic fibre suggestive of a pre-existing vessel wall.

Analysis at autopsy reveal that the majority of intracardiac varices is found in the right atrium, at the lower rim of the fossa ovalis and is thought to develop from dilated thrombosed veins. Of the reported patients, six were located in the right atrium, one located on the septal tricuspid leaflet (Ebstein’s anomaly), one located at the left ventricular outflow tract, another located on the mitral valve, and our case found in the left atrium. The inferior rim of the fossa ovalis is known to be the site of small veins, but the mechanism of how these veins dilate and form varices accompanied by calcification is still unclarified. One can only speculate that varices start off as cystic nodules, then develop phleboliths as a result of malperfusion, and with time, become filled with organizing thrombi.

The clinical importance and natural course of intracardiac varices is not well known, and surgical indications for asymptomatic intracardiac varices have not been made clear. However, by being aware of this entity, further reports may become apparent, and detailed analysis may unravel unique characteristics of this disease in the future. In the mean time, because of the difficulty in differentiation with other cardiac tumours, surgical resection according to the guidelines for benign intracardiac tumours may be a suitable strategy for intracardiac varices.

**Conflict of interest:** none declared.

**REFERENCES**