Intracardiac varices are very rare tumor-like conditions of the heart. They are endocardial, unilocular, blood-filled cysts lined by endothelial cells and filled with organizing thrombi. They are usually found in the right atrium, near the inferior rim of the fossa ovalis and develop from dilated thrombosed veins. The reported incidence is from 0.02% to 2.5%. Because they usually do not cause symptoms, most cases of intracardiac varices have been diagnosed at autopsy.1–3 Less than 6 cases have been diagnosed in living patients and most were preoperatively thought to be a myxoma.4–9

We report an unusual case of a 68-year-old woman who presented with an incidentally detected cardiac mass during ultrasonographic examination, which was confirmed as a varix of the heart by postoperative biopsy.

Case Report

A 68-year-old woman was transferred for evaluation of an incidentally detected asymptomatic cardiac mass. Imaging studies showed a 3.0×2.4 cm, well circumscribed, round, cystic mass with a calcified nodule that was attached to the lower rim of the fossa ovalis in the right atrium. Under cardiopulmonary bypass, the right atrium was opened to reveal a well circumscribed, dark bluish, pedunculated mass. Histologically, the specimen was a unilocular cyst lined by flattened endothelium, with peripheral fibrin clots and dystrophic calcification of the wall. Immunohistochemical staining of the lining cells was positive for cluster designation 34, which represents hematopoietic progenitor cell antigen. The final pathologic diagnosis was compatible with varix of the heart, which should be considered for a cystic mass with a calcified nodule located in the right atrium, near the lower rim of the fossa ovalis. (Circ J 2006; 70: 793–795)

Key Words: Cardiac mass; Varix
tricular systolic function. Computed tomography also showed a round cystic mass in the right atrium (Fig 1B).

Serologic tests for parasites (including Echinococcus) and tumor markers (including human chorionic gonadotropin) were negative. Thus, the most probable preoperative diagnosis of the mass was myxoma.

Under cardiopulmonary bypass, the right atrium was opened to reveal a well circumscribed, dark bluish to black, pedunculated mass attached to the lower rim of the fossa ovalis. After resection, the mass collapsed with spillage of dark venous blood (Fig 2A).

Macroscopically, the mass was a 2.8 cm, dark bluish cyst with a small pedunculated base. Histologically, it was a unilocular cyst lined by flattened endothelium, with peripheral fibrin clots and dystrophic calcification of the wall (Fig 2B). Immunohistochemical staining of the lining cells was positive for cluster designation 34, which represents hematopoietic progenitor cell antigen. The final pathologic diagnosis was compatible with varix of the heart.

The patient’s hospital course was uneventful and she was discharged on 14th hospital day.

**Discussion**

Primary tumors of the heart are rare, occurring at a frequency of 0.02% in a pooled autopsy series. Approximately three-quarters are histologically benign. Secondary or metastatic cardiac tumors occur more frequently, at approximately 100-fold the incidence of primary cardiac tumors.10,11

Intracardiac varices are very rare tumor-like conditions of the heart. They are endocardial, unilocular, blood-filled cysts lined by endothelial cells and filled with organizing thrombi. They are usually found in the right atrium, near the inferior rim of the fossa ovalis and develop from dilated thrombosed veins.1-3

Because intracardiac varices usually do not cause symptoms, most of the published cases have been diagnosed at autopsy. Heggtveit reported 25 cases of intracardiac varices in 1,000 consecutive autopsy studies of heart (2.5%); however, Rose reported only 4 cases in 5,887 autopsies (0.07%).2

Only less than 6 cases have been diagnosed in living patients. Thorsen et al first described a mobile intracardiac varix in a living patient with Ebstein’s anomaly, and it presented as a mobile mass on the septal leaflet of tricuspid valve detected by echocardiography that was confirmed as an intracardiac varix by pathologic examination. Harrity et al5 and Ramedi et al7 also reported intracardiac varices that were attached to the lower portion of right interatrial septum and confirmed by postoperative biopsy. The inferior rim of the fossa ovalis, where is the origin of almost all cardiac varices, is the site of small veins. In the present case, the varix was also located on the inferior part of the fossa ovalis.

The mechanism of these veins dilating and forming varices is uncertain. Heggtveit postulated that it may be embryonic incorporation of the remnants of the left valve of the sinus venosus into the right side of the interatrial septum. However, this explanation does not account for the rare intracardiac varices that occur in the ventricle. Salas Valien et al reported a case of a neonate who died of severe subaortic stenosis caused by a giant intracardiac varix of the left ventricular outflow tract.6 Although intracardiac varix usually does not cause clinical symptoms, it has important clinical significance in the differential diagnosis of other mass lesions in the right atrium. In most of the published cases, the preoperative diagnosis of the mass was myxoma, as in the present case also. Thus, the intracardiac varix needs to be differentiated from myxomas. Myxomas usually arise from the left atrium and only fewer than 20% arise in the right atrium; whereas varix of the heart arises in the right atrium, near the inferior rim of the fossa ovalis, and is an endocardial, unilocular, blood-filled cyst lined by endothelial cells. Cystic change of myxomas is an infrequent phenomenon. According to Acebo et al, cystic changes were noted in just 1 of 37 cases of cardiac myxomas.13 Varix of the heart is often associated with a densely calcified plebolith, whereas calcification of myxomas is infrequent. Acebo et al reported only 4 of 37 cases (11%) of cardiac myxomas with associated calcification.13 Based on these findings, varix of the heart could be differentiated from cardiac myxomas and therefore should be considered in the differential diagnosis of a cystic mass with calcified nodule located in right atrium, near the lower rim of the fossa ovalis.

Although it was usually asymptomatic, all reported cases including the present one underwent surgical removal of the mass. Because of its rarity there are no guidelines for the surgical indication for asymptomatic intracardiac varix. Thus, it seems reasonable to choose the treatment according to the guidelines of the other benign cardiac masses, such as hemodynamic derangement, or risk of embolization.

**References**


