

Original Article

Extremely rare case of primary cardiac chondroma in a patient presenting with acute pulmonary edema

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Abstract

Primary tumors of the heart are rare entities, substantially less frequent than cardiac metastases. Primary chondroma is an exceptionally rare cardiac tumor. We describe the case of a patient presenting with acute pulmonary edema with the incidental echocardiographic finding of a large left atrial tumor that was histopathologically diagnosed as primary cardiac chondroma. © 2011 Elsevier Inc. All rights reserved.

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A 62-year-old man with a history of myocardial infarction and coronary revascularization was referred to our hospital with 3-day onset of exertional dyspnea and chest pain. The electrocardiogram showed ST-segment depression in the anterior leads. Biochemical tests including cardiac enzymes were within normal limits. Shortly after admission, the patient developed acute pulmonary edema.

Transthoracic echocardiogram demonstrated a heterogeneous mass with predominant echodensity and regions with cystic appearance, within the left atrium (Fig. 1A). The mass protruded into the left ventricle during diastole, obstructing the mitral valve orifice (Fig. 1B). No additional imaging test was performed due to the acute clinical deterioration. The patient was urgently referred for tumor excision. The mass was arising from the left atrial wall through a narrow stalk adjacent to the posterior mitral valve annulus and did not invade the myocardium. Macroscopically it was smooth,

multi-lobulated, predominantly solid, and measured 6.1×3.9 cm (Fig. 2). Histological examination from multiple sections showed pure benign hyaline chondroid tissue with regions of cystic degeneration and absence of cytologic atypia, abnormal mitotic figures, sarcomatous, or myxoid regions (Fig. 3). No further chondroid lesions were identified at the time of surgery. Postoperative clinical evaluation and imaging testing excluded potential extracardiac tumors with chondroid differentiation, such as teratoma, and primary cardiac chondroma was diagnosed. The patient had an uneventful postoperative course, with a normal echocardiogram at 3-month follow-up (Fig. 1C, D).

Primary cardiac tumors are rare entities, 20–40 times less common than cardiac metastases, with a prevalence ranging between 0.001% and 0.25% [1]. They may be asymptomatic or display nonspecific clinical manifestations including obstructive and embolic complications, depending on their size, location, and mobility. Soft tissue chondromas occur at various extraskelatal anatomic locations; visceral chondromas are typically localized in the lungs, often as a manifestation of Carney's syndrome [2]. Cardiac chondroma is an extremely rare primary cardiac tumor, with only two cases previously reported to our knowledge [3,4]; hence the natural history and prognosis of this tumor are unknown.

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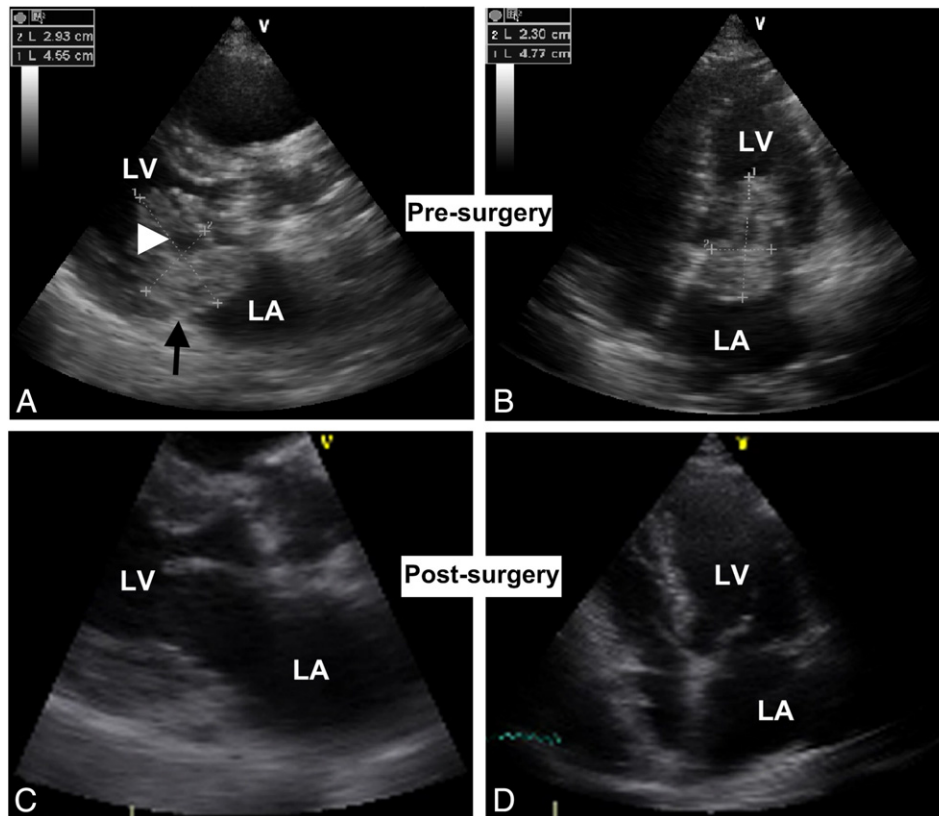


Fig. 1. (A,B) Preoperative transthoracic echocardiogram. Parasternal long axis view (A) shows a heterogeneous, predominantly echodense left atrial mass with focal cystic appearance (white arrowhead) attached to the left atrial wall through a narrow stalk (black arrow). (B) Apical four-chamber view shows the mass projecting into the left ventricle during diastole. (C,D) Follow-up echocardiogram. Parasternal long axis (C) and apical four-chamber view (D) show normal appearance of the left atrial cavity. LA indicates left atrium; LV indicates left ventricle.

The chondroid differentiation in this tumor is of unspecified origin, since the human heart does not normally contain cartilage. Chondrocytes have been implicated in valvular myxomatous degeneration secondary to an osteoblast differentiation and endochondral bone formation process in mitral valves [5]; however, the mass was not arising from the mitral valve in our patient. Other cartilage-containing cardiac tumors include primary or metastatic chondrosarcoma or teratoma, and myxoma with focal chondroid differentiation, which were histopathologically excluded in our case.

A high index of suspicion is required in patients presenting with acute onset dyspnea, to ensure that the diagnosis of an underlying cardiac mass is not missed. Although extremely rare, atrial chondroma should be considered in the differential diagnosis of cardiac space-occupying lesions. Despite its histologically benign nature, cardiac chondroma may be life-threatening because of its strategic position, unless promptly identified and excised.

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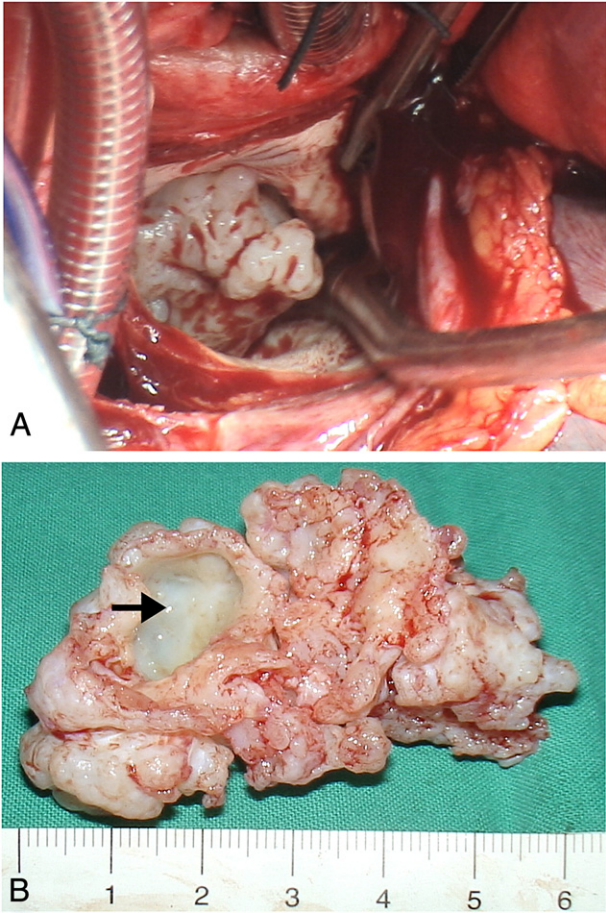


Fig. 2. (A) Intraoperative image from right thoracotomy showing the occupation of the left atrium by the tumor. (B) Macroscopic photograph illustrating the white, multi-lobulated external appearance and the cystic degeneration (arrow) of the excised tumor, measuring 6.1×3.9 cm.

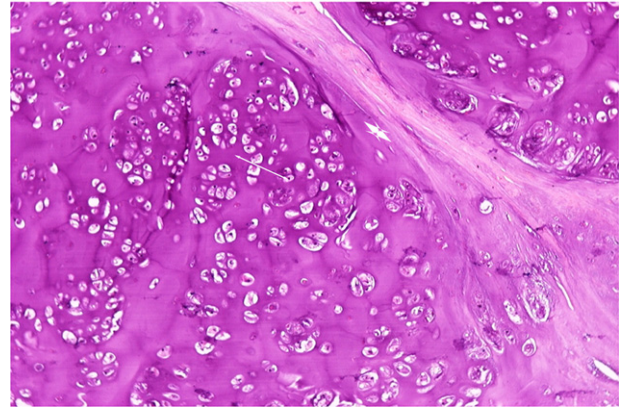


Fig. 3. Photomicrograph of the tumor demonstrating the hyaline chondroid tissue. Hematoxylin–eosin staining, magnification ×10.