



A Calcified Amorphous Tumor in the Left Atrium: A Case Report

좌심방에서 발생한 석회화 무정형 종양: 증례 보고

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Calcified amorphous tumors (CATs) of the heart are rare non-neoplastic cardiac masses primarily found in the mitral valve or annulus. However, their exact pathogenesis remains unknown. In this case report, we describe the CT and MRI findings and differentiating features of cardiac a CAT in the left atrium of a 79-year-old female.

Index terms Case Report; Calcified Amorphous Tumor; Left Atrium; Cardiac Calcified Lesion

INTRODUCTION

Cardiac calcified amorphous tumors (CATs) are rare non-neoplastic cardiac masses with microscopic calcification features and amorphous fibrinous material (1). An association between CATs, mitral annular calcification (MAC), and end-stage renal disease (ESRD) has been reported. Cardiac CATs may present with symptoms such as shortness of breath and systemic embolization (1). Cardiac CATs in the left atrium (LA) and interatrial septum are rare. To our knowledge, only six such cases have been reported (2-4). Only a few case reports have also described cardiac the CT and MRI findings of CATs. Here, we report the case of a 79-year-old female with a cardiac CAT in the LA and describe the multimodality imaging and pathologic findings.

CASE REPORT

A 79-year-old female was incidentally found with a cardiac mass on chest CT during a health checkup. The patient had undergone surgery for colon cancer and had diabetes and hearing loss. She was neither an alcoholic nor a smoker. Her serum

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calcium level and kidney function were within normal ranges. She had no other abnormalities in laboratory findings. A health check-up with a non-contrast-enhanced chest CT showed a 2-cm calcified mass in the LA (Fig. 1A). Cardiac MRI showed a 2 cm-sized mass in the LA. Cine imaging revealed that the mass was attached to the interatrial septum of the LA (Fig. 1B,

Fig. 1. A calcified amorphous tumor at the interatrial septum of the LA in a 79-year-old female.

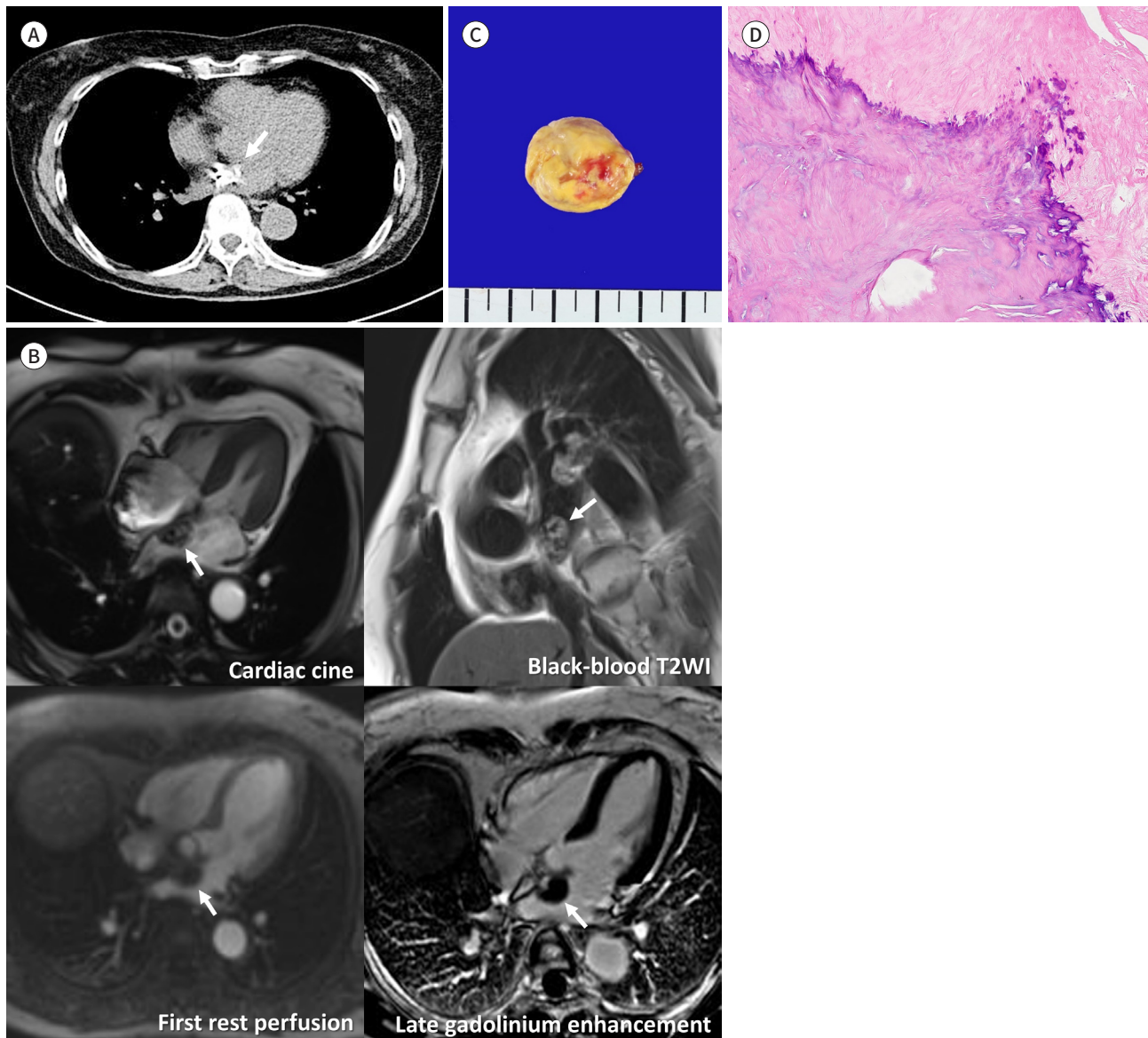
A. The initial low-dose non-contrast enhanced chest CT image shows a 2 cm-sized calcified mass (arrow) in the LA attached to the interatrial septum.

B. Cardiac cine image shows that the low signal intensity mass attaches at the LA's interatrial septum (arrow). On the black-blood T2-WI image, the mass (arrow) shows intermediate signal intensity with low signal intense foci. The mass (arrow) shows no enhancement on the first pass rest perfusion image. On late gadolinium enhancement, the mass (arrow) shows no enhancement.

C. The mass grossly appears as a well-defined yellowish tumor.

D. Microscopic findings reveals mostly acellular hyalinized stroma and dystrophic calcification. The stroma alternates between dense and loose areas. There are few inflammatory cells, and no definite neoplastic cells or micro-organisms are seen (hematoxylin and eosin stain, $\times 100$).

LA = left atrium, WI = weighted image



Supplementary Video 1A). The mass showed heterogeneous high signal intensity (SI) on T2-weighted imaging (WI; Fig. 1B) and no enhancement on first-pass rest perfusion (Fig. 1B, Supplementary Video 1B) or late gadolinium enhancement (LGE) (Fig. 1B) sequences. Owing to the potential risk of thromboembolism, surgical removal of the LA mass and interatrial septum patch repair were performed. Microscopically, the mass was primarily composed of hyalinized stroma with degeneration, accompanied by dystrophic calcification (Fig. 1D). No myxomatous components or malignant cells were observed. The histopathological diagnosis was a cardiac CAT. The patient recovered without any complications and was discharged. The patient is currently undergoing outpatient follow-up. This study was approved by the Institutional Review Board of our institution, which waived the requirement for informed consent (IRB No. 2023-10-004).

DISCUSSION

Primary intracardiac tumors are uncommon, accounting for only 0.2% of all tumors. Among adults, cardiac myxomas are the most frequently diagnosed benign primary intracardiac tumors, constituting approximately 20%–30% of cases. A cardiac CAT is a rare non-neoplastic cardiac mass microscopically composed of calcified nodules in an amorphous fibrin background with degeneration and focal inflammation (1). However, the exact pathogenesis remains unknown. It may be associated with organized thrombi, primary or secondary hypercoagulability, or abnormal calcium and phosphorus metabolism, particularly in patients on hemodialysis (5). Previous studies have reported that cardiac CATs are often associated with valve disease, MAC, ESRD, diabetes, and coronary artery disease (1). Patients with CATs may be asymptomatic or present with symptoms related to obstruction or embolization, such as shortness of breath, syncope, and stroke (1, 6). Cardiac CATs were found in all the cardiac chambers, predominately in the mitral valve or annulus, followed by the left ventricle and right atrium. To our knowledge, only 95 cardiac CAT cases have been reported (Supplementary Table 1). They are rare in the LA and interatrial septum; only six cases have been reported (2-4).

As in this case, cardiac CATs are mostly seen as partially calcified hypodense masses or diffuse calcified masses without enhancement on CT scans (6). A previous study reported that cardiac CATs typically exhibit homogeneous low SI on T1- and T2-WI and do not show enhancement after administering gadolinium contrast agents in early and delayed sequences (6). In our case, the cardiac CAT showed heterogeneously high SI on T2-WI. This is because calcifications can show various SI on MRI. The configuration and shape of cardiac CATs vary in all published cases. The size of the masses varies between 1.3 cm and 4.3 cm in diameter (6). Our cardiac CAT case was found in a rare location, the interatrial septum of the LA, and the patient did not have ESRD, coronary artery disease, heart valve disease, or MAC. However, this patient had a history of diabetes, which is associated with CAT. Although patients with diabetes and ESRD have a higher prevalence of CATs, the exact mechanism has not yet been elucidated. Cardiac CATs should be differentially diagnosed from other cardiac tumors, such as calcified myxoma, organized calcified thrombus, cardiac osteosarcoma, and cardiac fibroma, for appropriate treatment.

Cardiac myxomas are the most common primary cardiac tumors and are most commonly

found in the LA, attached by a stalk to the fossa ovalis (7). On CT, myxomas typically manifest as low-attenuation masses with smooth or slightly villous surfaces. Arterial-phase contrast enhancement is usually absent; however, delayed heterogeneous enhancement may be observed on CT. Calcification is seen in approximately 14% of myxomas, and most show punctate or eccentric forms of calcium (7). On cardiac MRI, myxomas show heterogeneous SI on T1- and T2-WI, varying according to the amount of myxoid, hemorrhagic, ossific, and necrotic tissue components. The most important distinguishing feature is that myxomas show patchy delayed gadolinium enhancement (7).

Cardiac fibromas may also exhibit intratumoral calcification. However, they are primarily observed in the pediatric population and typically arise in the ventricular walls. Calcification was observed in approximately 25% of cases, but most were punctate. Mural location is a distinguishing feature of cardiac fibromas that differentiates them from other cardiac tumors (8). On CT, cardiac fibromas typically show homogeneous or heterogeneous enhancement after the administration of an intravenous contrast material. On MRI, cardiac fibromas show iso-SI relative to the myocardium on T1-WI and low SI on T2-WI. In contrast to cardiac CATs, cardiac fibromas generally exhibit intensely delayed hyperenhancement (8).

Caseous MAC is a rare variant of MAC, mainly located in the posterior annular region of the mitral valve. Caseous MAC generally appears as a round, sometimes semilunar, mass with peripheral calcification and central liquefaction necrosis and lacks contrast enhancement on CT (9). MRI may show an enhanced peripheral border surrounding a non-enhanced core (9).

Primary osteosarcomas predominantly occur in the LA and often exhibit dense calcification. Because of their location and calcification, they can also be included in the differential diagnosis. However, the tumor's broad attachment base and invasive features should be suspected. It appears as a heterogeneously low SI on T1-WI, high SI on T2-WI, and avid enhancement (10).

We report a rare case of a cardiac CAT in the interatrial septum of the LA. This case report presents the differential diagnosis of a calcified atrial mass and differentiating imaging findings. In cases of extensively calcified tumors that do not exhibit enhancement, CATs should be considered in the differential diagnosis.

Supplementary Materials

The Supplement is available with this article at <http://doi.org/10.3348/jksr.2023.0133>.

Supplementary Video Legend

Video 1. Cardiac MRI of a calcified amorphous tumor at the interatrial septum of the left atrium (LA) in a 79-year-old female.

- A. The cine image shows a mass attached to the interatrial septum.
- B. The rest first-pass perfusion image shows no enhancement of the mass.






Author Contributions

Conceptualization, K.J.Y., P.N.H., K.Y.S.; data curation, K.M.S., K.J.Y., H.J.H., L.H.W., P.N.H.; investigation, K.M.S., K.J.Y., L.M.S., L.H.W.; methodology, K.J.Y., L.H.W.; project administration, K.J.Y.; supervision, K.J.Y., H.J.H., P.N.H.; validation, L.M.S., K.Y.S.; visualization, K.J.Y., K.Y.S.; writing—original draft, all authors; and writing—review & editing, K.M.S., K.J.Y., L.M.S., H.J.H.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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좌심방에서 발생한 석회화 무정형 종양: 증례 보고

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심장에 발생하는 무정형 석회화 종양은 드문 비종양성 종괴로 승모판막 또는 승모판막 고리에 가장 흔하게 발생하며 정확한 발생 원인은 알려져 있지 않다. 본 증례 보고에서는 79세 여성의 좌심방 심방중격에 발생한 무정형 석회화 종양의 CT와 MRI 영상 소견과 감별진단에 관하여 이야기하고자 한다.

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