Idiopathic cardiac ossification with Chiari mesh in the right atrium: a case report and literature review

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Background: Idiopathic cardiac osseous metaplasia in the right atrium of a 9-year-old boy, accompanied by right atrial Chiari network and right pulmonary artery embolism. This case is rare and can easily be misdiagnosed.

Case Description: We encountered a case of a 9-year-old boy with a 3.5 cm diameter neoplasm in the right atrium. Preoperative imaging diagnosis could not determine the nature of the tumor, and the initial clinical suspicion of cardiac myxoma. After admission, a cardiotomy to remove foreign bodies and a pulmonary artery thrombectomy were performed.

Conclusions: Idiopathic cardiac osseous metaplasia is relatively rare, and it is even rarer to be accompanied by a Chiari network in the right atrium. Due to the location and characteristics of the lesion in this case, it is easy to be misdiagnosed as atrial myxoma in clinical practice. Whether it is idiopathic osseous metaplasia or myxoma, it needs to be performed surgical treatment and pathological examination can easily rule out the diagnosis of myxoma. However, as idiopathic cardiac metaplasia is difficult to encounter in clinical work and there are few reports in the literature, clinicians and pathologists need to consult more relevant literature. Learn to understand and master the disease through multi-party consultation.

Keywords: Idiopathic cardiac ossification; Chiari mesh; right atrium; case report

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Introduction

Myocardial calcification is an unusual entity. Scholz *et al.* were the first to radiologically demonstrate myocardial calcification in 1919 (1). Since then, there have been various reports of myocardial calcification, most commonly in men and usually associated with either coronary artery disease or a history of myocardial damage. The heart may calcify at different locations, such as valves, arteries, endocardial, pericardium, and conduction tissue (2,3). Degeneration and inflammatory mechanisms have been suggested as possible explanations for these calcifications. Intramuscular calcification is extremely rare and is associated with chronic renal failure (especially hemodialysis treatment) and secondary hyperparathyroidism. We present this case in accordance with the CARE reporting checklist (available at https://acr.amegroups.com/article/view/10.21037/acr-23-168/rc).

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's parents or legal guardians

for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

General information

The patient is a 9-year-old boy. Due to sudden syncope after physical education class, he regained self-awareness after about 2 minutes and went to see a doctor. No convulsions, vomiting, or incontinence occurred during the onset. There is no special medical history or family history. The patient had suffered from new coronary pneumonia half a month before the onset of the illness. The initial clinical diagnosis was suspected to be myxoma in the right atrium. After admission, a "cardiotomy to remove foreign bodies and pulmonary artery thrombectomy" was performed. During the operation, it was found that the vegetation was located at the point where the inferior vena cava enters the right atrium. It was cauliflower-shaped and about 3.5 cm × 2.5 cm in size. The texture on the side near the right atrium is soft, and the texture on the inferior vena cava side is hard, calcified, and yellow. The root is adherent to the inferior vena cava valve and is closely related. Exploration of the right pulmonary artery revealed that a thrombus-like substance almost completely blocked the right pulmonary artery, about 2.5 cm × 1.5 cm in size, soft, yellow, and branch-like.

Highlight box

Key findings

- This report describes a case that idiopathic cardiac osseous metaplasia in the right atrium of a 9-year-old boy, accompanied by right atrial Chiari network and right pulmonary artery embolism.

What is known and what is new?

- Idiopathic cardiac osseous metaplasia is relatively rare, and it is even rarer to be accompanied by a Chiari network in the right atrium.
- Due to the location and characteristics of the lesion in this case, it is easy to be misdiagnosed as atrial myxoma in clinical practice.

What is the implication, and what should change now?

- The diagnosis of idiopathic cardiac metaplasia ultimately relies on pathological examination, but imaging results are important for clinical evaluation before surgery. The doctor has a certain prompting effect. Immunohistochemical staining and special staining have limited auxiliary roles in the diagnosis of this disease. The diagnosis of idiopathic cardiac osseous metaplasia mainly relies on the diagnosis of histomorphology by pathological gross examination and conventional microscopic staining.

Imaging examination

Cardiac ultrasound, computed tomography (CT), and maximum intensity projection (MIP) suggest: enlargement of the right heart, strong echo group where the inferior vena cava enters the right atrium orifice, suspicious thrombus, band-like strong echo in the right atrium, consider Chiari mesh, hypoechoic was detected on the right atrial side of the fossa ovale, the tricuspid valve had moderate to severe regurgitation, and pulmonary hypertension (*Figure 1A-1D*); pulmonary artery CT showed multiple embolisms in the main right pulmonary artery and branches of both pulmonary arteries.

Pathological examination

General specimen examination: foreign body in the heart is several pieces of grayish-white soft tissue, 1.3 cm × 1.5 cm × 0.5 cm in size, its surface is rough grayish-white, grayish-yellow on the cut surface, and it is brittle; also pathologist have seen a piece of hard tissue, 2.5 cm × 2 cm × 1.7 cm in size, the surface is multi-nodular, grayish-pink, grayish-yellow and rough, the cut surface is solid (*Figure 2*); (pulmonary artery foreign body and thrombus are a piece of gray-red soft tissue, 2.8 cm × 1.7 cm × 0.6 cm in size, gray-pink, gray-red and rough on the surface, gray-pink, gray-red on the cut surface, soft, partial very crispy (*Figure 3*).

Histopathological morphology was examined under the microscope: (foreign body in the heart) microscopic examination shows that it is composed of mature bone tissue, bone marrow tissue, and fibrous tissue, accompanied by calcification and necrosis (*Figures 4,5*); (foreign body in the pulmonary artery and thrombus) platelets aggregate into trabeculae. The spaces are filled with a large amount of coagulated fibrin and red blood cells and white blood cells can be seen at the edges of the trabeculae.

Immunohistochemical staining: vimentin (+), AE1/AE3 (–), desmin (+), SMA (+), CD117 (scattered +), CD34 (vascular +), Ki-67 (<1%). Special dyeing: mesh fiber, periodic acid-Schiff (PAS), hexamine silver.

Pathological diagnosis: (cardiac foreign body) idiopathic ossification; (pulmonary artery foreign body and thrombus) mixed thrombus.

Follow-up

After 5 months of follow-up, the patient was in good

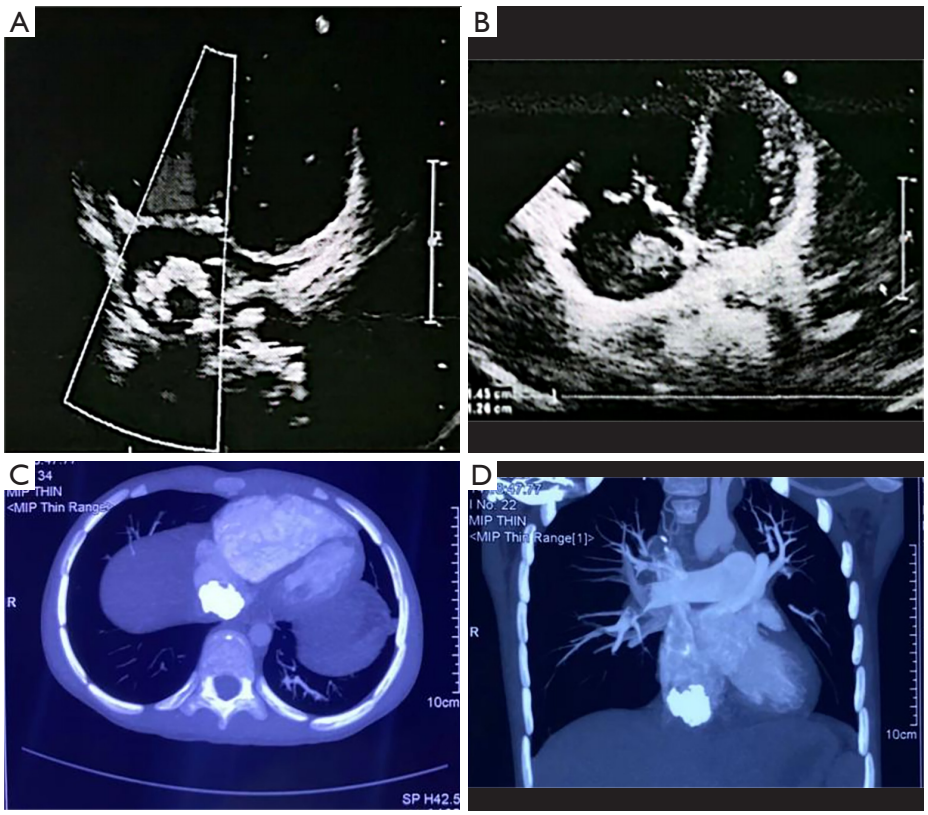

Figure 1 There was a hyperechoic mass with a size of 24.9 mm × 7.7 mm where the inferior vena cava entered the right atrium, and a hypoechoic mass with a size of 15.5 mm × 12.6 mm on the right atrial side of the fossa ovale, which seemed to be connected with the fossa ovale by a pedicle. (A,B) Echocardiogram. (C) Cardiac-enhanced CT. (D) Cardiac MIP. MIP, maximum intensity projection; R, right; CT, computed tomography.

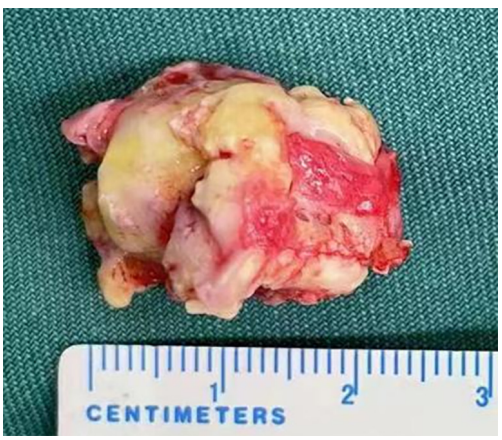

Figure 2 Heart disease, cauliflower-like appearance, rough grayish-pink surface, grayish-pink grayish-yellow cut surface, and brittle texture.

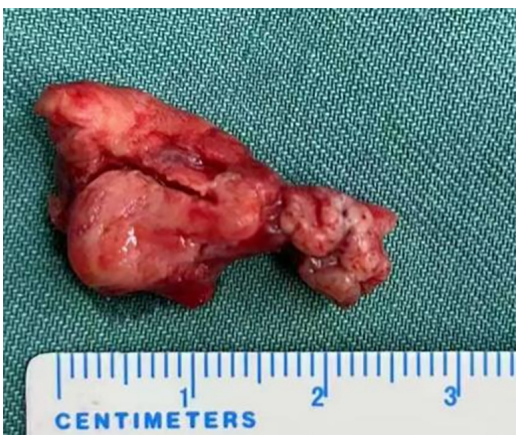

Figure 3 Pulmonary artery disease, the surface is gray-pink, gray-red, and rough, the cut surface is gray-pink, gray-red, and soft, and some parts are brittle.

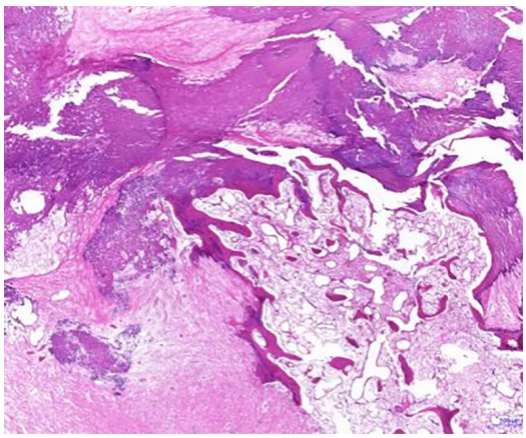

Figure 4 HE staining low magnification 100×: composition of bone tissue, bone trabeculae, bone marrow tissue, and fibrous tissue, accompanied by calcification and necrosis. HE, hematoxylin-eosin.

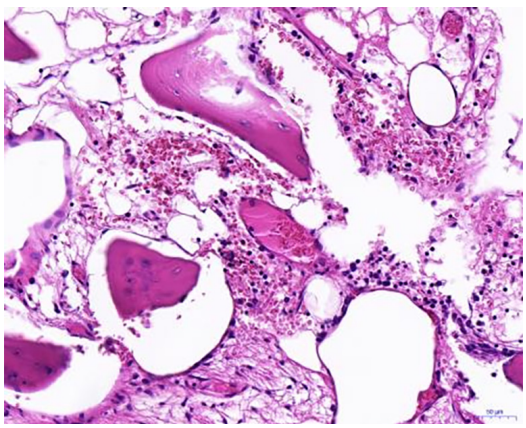

Figure 5 HE staining high magnification 200×: mature bone tissue, bone marrow tissue, and fibrous tissue. HE, hematoxylin-eosin.

condition.

Discussion

Pathologically, calcification is divided into dystrophic calcification and metastatic calcification. In the cardiovascular system, dystrophic calcification is often seen in the coronary arteries, mitral annulus, and aortic valve due to tissue degeneration. However, cardiac calcification with bony metaplasia is uncommon, and even rarer in children.

As early as 1919, Scholz observed calcified heart on X-ray films (1). Catellier reported four cases of cardiac calcification deposition in the literature. Three of the patients were middle-aged and elderly, and all had hypertension and end-stage renal disease. One of them was accompanied by hyperparathyroidism, and the other was a 6-year-old boy suffering from patients with congenital cyanotic heart disease, damage caused by extensive surgical repair, damage caused by congenital heart defects, and bacterial myocarditis can all lead to dystrophic calcification of the heart in patients. Calcification sites include the mitral valve, aortic valve, pulmonary valve, left atrium, left ventricle, and interventricular septum, etc. The locations are widespread and may be related to calcium salt metabolism or myocardial inflammation (2,3). Myocardial calcification has also been reported after septic shock. The author proposed that the cause of calcium deposition is catecholamine-induced myocardial cell necrosis, which leads to increased membrane permeability and calcium ion influx (4). There is also a report of a female patient without metabolic disease, trauma history, or other diseases, but severe calcification was found in the left ventricular myocardial tissue (1). El-Bialy *et al.* also reported a similar case of this type of calcification. The patient was an 80-year-old woman who developed severe myocardial calcification within 4 years, and the etiology could not be determined (5).

The patient in this case was a 9-year-old boy. He had no history of metabolic diseases or family diseases before the onset of the disease. The lesion was located at the mouth of the inferior vena cava entering the right atrium. The age and location of the onset were relatively rare, and the pathogenesis was unclear. In this patient, bone, bone marrow tissue, and fibrous tissue were visible in the calcified lesions. Literature reports indicate that this type of disease is called idiopathic osseous metaplasia (6). Studies have shown that key factors for the development of heterotopic ossification in humans include hypoxia, a high degree of injury, or prolonged inflammatory response (7).

The cardiac ultrasound in this case showed a band-like strong echo in the right atrium, with one end connected to the coronary sinus and one end connected to the inferior vena cava into the right atrium, close to the location of the mass. Considering the Chiari network, this may be related to the calcification ossification and mixed pulmonary artery thrombus that occurred in this patient at the opening of the inferior vena cava into the right atrium. Chiari nets are relatively rare, with an incidence of about 2% in the general population (8). First described by Hans Chiari at the end of the 19th century, it is one of the embryological

remnants of the right venous sinus valve (9). Studies have shown that the Chiari network is related to cardiovascular complications, such as thromboembolism, supraventricular arrhythmia, atrial septal defect, or cyanosis (10,11). The statement that Chiari net causes thromboembolic disease is still controversial. On the one hand, Chiari net may cause atrial thrombosis due to its impact on the hemodynamics of the right atrium, making the blood flow easily to the interatrial septum, causing turbulence, thereby forming a thrombus. The net can intercept the fragments of the thrombus and form a larger thrombus. Once the thrombus gradually increases or becomes unstable, it may lead to a fatal pulmonary embolism (12). On the other hand, the Chiari net may become a protective filter against pulmonary embolism by intercepting thrombus (13). This patient had multiple embolisms in the main right pulmonary artery and branches of both pulmonary arteries. Pathology confirmed mixed thrombi. It was considered that calcification and ossification occurred at the inlet of the right atrium. In addition, the presence of the Chiari network in the right atrium made the blood flow more prone to turbulence and thrombosis. It falls off and becomes blocked as the blood flows into the pulmonary artery. Thrombus can also be seen under the microscope at the entrance of the right atrium, which can confirm that pulmonary embolism is caused by the falling off of the thrombus at the heart lesion. Other studies have shown that chronic right atrium thrombus can sometimes lead to surrounding tissue destruction (14).

CT plays an important role in diagnosing myocardial calcification before surgery and is highly sensitive to the location and nature of calcification (5). The diagnosis of idiopathic cardiac metaplasia ultimately relies on pathological examination, but imaging results are important for clinical evaluation before surgery. The doctor has a certain prompting effect. Immunohistochemical staining and special staining have limited auxiliary roles in the diagnosis of this disease. The diagnosis of idiopathic cardiac osseous metaplasia mainly relies on the diagnosis of histomorphology by pathological gross examination and conventional microscopic staining.

Since the preoperative diagnosis of this patient was not clear, surgery was performed, and postoperative pathological results confirmed idiopathic cardiac ossification. Patients with idiopathic cardiac ossification mostly occurred in the myocardium, suffered from severe heart failure, and most of them underwent heart transplantation. However, due to severe inferior vena cava obstruction during surgical treatment, inferior vena cava cannulation could not be completed, which brought certain difficulties and challenges to cardiopulmonary bypass heart surgery. In the end, the diseased tissue was completely removed by surgery, and the operation was successfully completed. All the indicators of the postoperative examination were normal, and the reexamination at this stage was also normal.

Conclusions

Idiopathic cardiac osseous metaplasia is relatively rare, and it is even rarer to be accompanied by a Chiari network in the right atrium. Due to the location and characteristics of the lesion in this case, it is easy to be misdiagnosed as atrial myxoma in clinical practice. Whether it is idiopathic osseous metaplasia or myxoma, it needs to be performed Surgical treatment and pathological examination can easily rule out the diagnosis of myxoma.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://acr.amegroups.com/article/view/10.21037/acr-23-168/rc

Peer Review File: Available at https://acr.amegroups.com/article/view/10.21037/acr-23-168/prf

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://acr.amegroups.com/article/view/10.21037/acr-23-168/coif). The authors report that the study was supported by the Beijing Hospitals Authority InnovationStudio of Young Staff Funding Support (No. 202114). The authors have no other conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures

performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's parents or legal guardians for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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