Atypical localization, atypical clinical course, unexpected age: Myxoma

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Introduction

The incidence of cardiac tumors in infants and the pediatric population is 0.08% (1). Rhabdomyoma is the most common tumor in the neonatal period (1, 2). However, myxoma is the most common primary tumor that is frequently localized in the left atrium in the adult population. The incidence of myxoma in the pediatric population is 20.2%. In fact, myxoma equally affects both atria during the pediatric ages (3).

Case Report

Our patient was born at the age of 32 weeks via elective cesarean section. She was followed-up in the neonatal intensive care unit for one month because of prematurity complications. After one month without any symptoms after discharge, she was

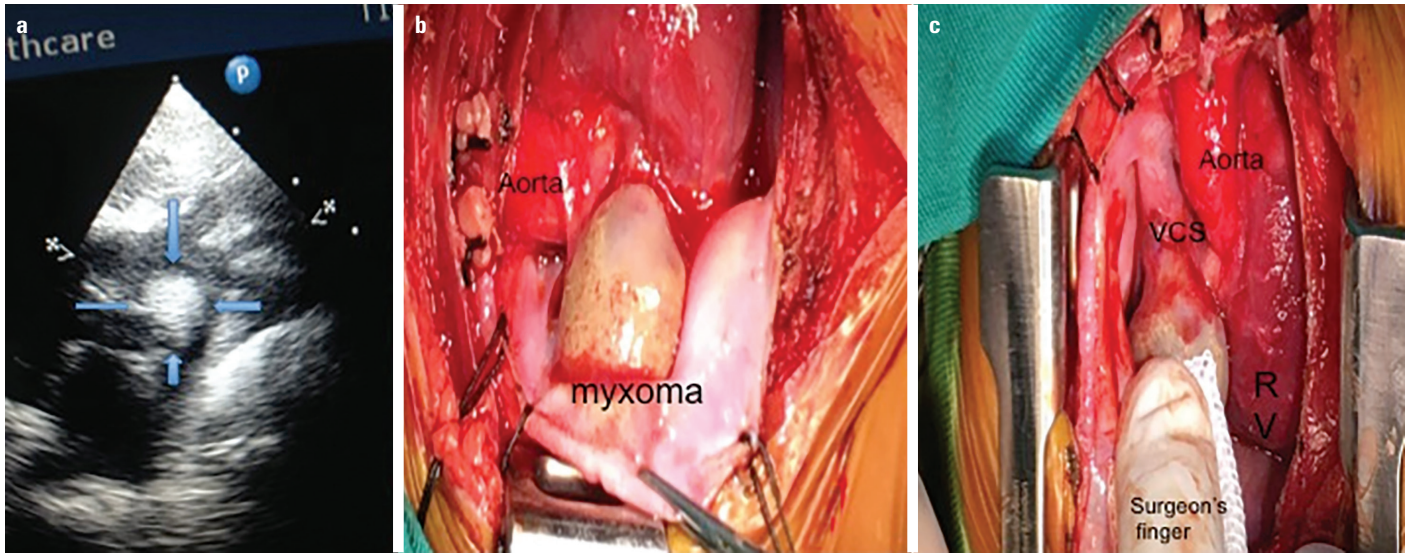

Figure 1. (a) Preoperative echocardiographic view of the cardiac myxoma. (b) The myxoma originating from the right atrium. (c) The myxoma adjacent to the sinus node and right coronary artery

admitted to our hospital with dyspnea. It was not found cyanosis or embolism of the extremities. The echocardiogram showed a massive pericardial effusion and a mass next to the right atrium. Additionally, 50 cc of hemorrhagic effusion was obtained via pericardiocentesis. Control echocardiography revealed that the mass was 2×3 cm in size, homogeneous, spherical, hyperechoic and located next to the right atrium, but there was no evidence of compression (Fig. 1a). No additional congenital heart anomaly was found. Under indomethacin treatment, massive effusion reoccurred and two additional pericardiocentesis were performed. No infectious agent in the effusion material was found.

The mass was explored via median sternotomy. It originated from the right atrial appendix, protruding externally without any peduncle formation. It was solid, avascular, ovoid, yellowish and 2×3×3 cm in size (Fig. 1b). Because of the localization near the sinus node and right coronary artery (Fig. 1c), we decided to resect it by using cardiopulmonary bypass. Aortic and selective venous cannulations were performed; after the cardiac arrest at 34°C, the mass was completely resected without any damage to the adjacent structures. There was no thrombus in the right atrium and the cavity was adequate. Thereafter, the atrium was closed using primary suturing (Fig. 2a). The cross-clamping time was 15 minutes and spontaneous sinus rhythm was obtained. The bypass duration was 45 minutes. The patient was extubated postoperatively after three hours, and she did not require any inotropic agent.

Macroscopic pathological examination revealed that the mass was semi-hard, gelatinous, friable and yellow–brown in color (Fig. 2b, 2c). Additionally, microscopic examination found that the myxoma cells were surrounded by myxoid stroma and collagen-rich stroma material. There were no atypical cells, pleomorphism or atypical mitose. The immunohistochemistry of desmin was negative (Fig. 2d). During the clinical course, there was neither pericardial effusion nor arrhythmia. After a month, there was no occurrence of pericardial effusion (Fig. 2e).

Discussion

Most cardiac masses can be detected via fetal echocardiography. Adverse hemodynamic effects can be easily observed with a close follow-up. In this case, the mother did not receive a suitable obstetric follow-up. Additionally, the asymptomatic baby was not examined while being in the neonatal care unit. Therefore, the diagnosis was discovered with the presentation of cardiac tamponade. In the neonatal period, every resectable cardiac mass that is still causing symptoms should be resected (3). Primary intracardiac tumors in the neonatal period are rhabdomyoma, fibroma, myxoma, teratoma and hemangioma (2). Our patient had cardiac myxoma, which is very rare in childhood but frequent in adult patients. For all ages, myxoma generally originates from the interatrial septum but can be present in every heart chamber, especially in the left atrium. There may be an occurrence of blood flow obstruction and embolic processes. Occasionally, extracardiac myxomas are observed in the right ventricular epicardium, superior or inferior vena cava, femoral vein, as well as in head and neck as odontologic tumors. In very rare cases, intracardiac localization is observed in the Eustachian valve (4, 5). Myxomas are semisolid masses rather than fibromas (2).

In our case, although the myxoma originated from the right atrial appendix and did not compress any cardiac structure, it was too large for a low-birth-weight baby. The indications for surgery were the life-threatening cardiac tamponade and recurrent pericardial effusion that were refractory to medical and interventional therapies.

Our female patient confirms that there is a higher prevalence of myxoma in females.

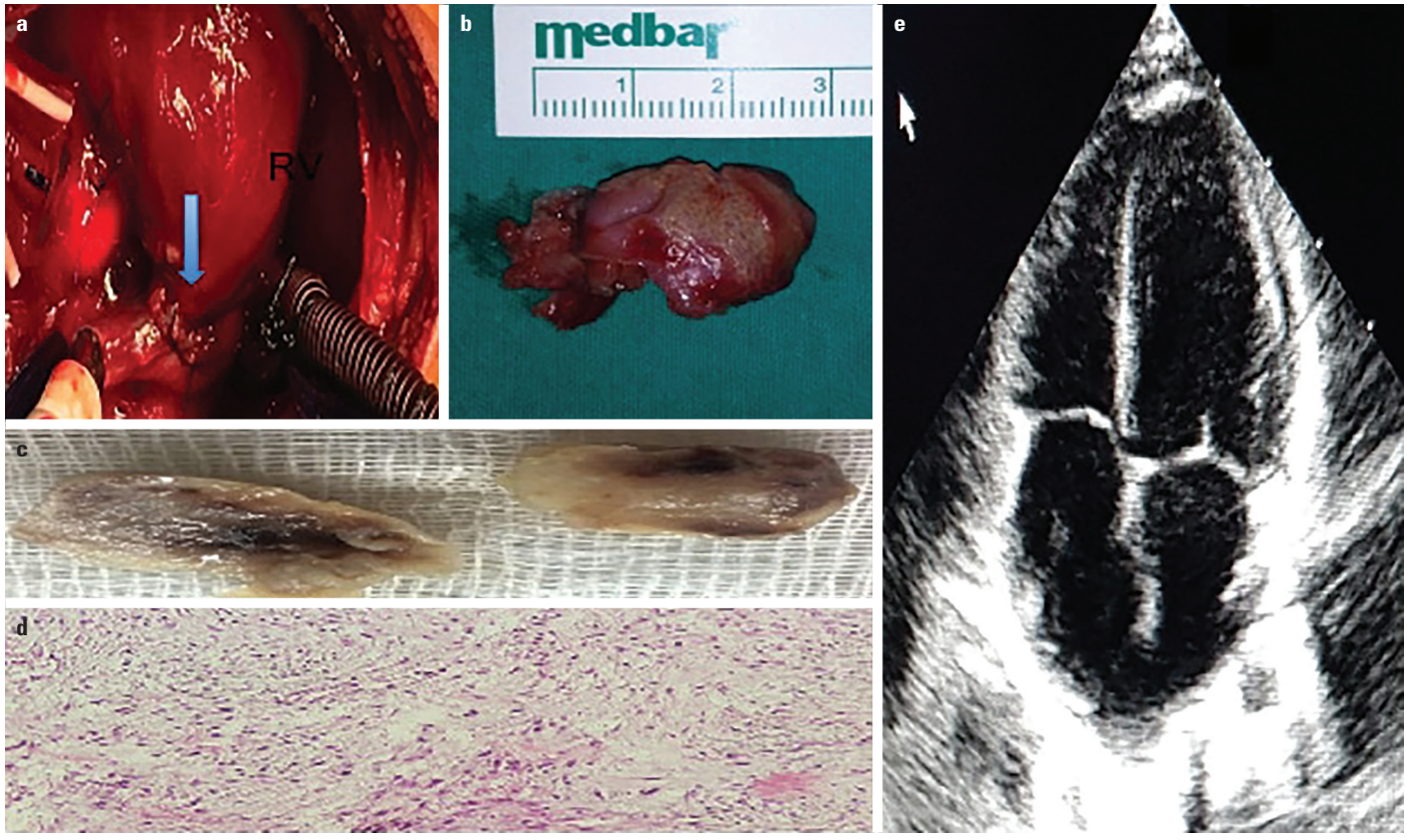

Figure 2. (a) Right atrium after the resection of the myxoma. (b) Macroscopic view. (c) Gelatinous and friable features. (d) Microscopy with hematoxylin and eosin staining. (e) Echocardiography on postoperative day 30

As far as we know, our patient is the second in the literature to undergo surgery at the weight of 2.75 kg with cardiopulmonary bypass for myxoma unexpectedly originating from the right atrial appendix. Besides her low weight, at the corrected age of 40 weeks, she is the second youngest patient to undergo surgery for myxoma (6).

Conclusion

After the diagnosis of the unexpected mixoma in a low-birth-weight infant, resection of the tumor, close to the important structures as right coronary artery and the sinus node, was challenging. To aviod the damage and postoperative complications in infants, the resection of tumors as such in our case, should be performed using cardiopulmonary bypass process.

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