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Obstructive neonatal atrial myxoma

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ABSTRACT

INTRODUCTION: Cardiac myxoma is a benign neoplasm representing the most common primary cardiac tumor in adults, however it is unusual in neonates. It is represented by an endocardial mass that occupies the cardiac chamber. Although the majority of myxomas are attached to the fossa ovalis of the interatrial septum, they also attach to the walls of the cardiac chambers and to valve leaflets surfaces. Approximately 75% of myxomas are found in the left atrium, 20% are located in the right atrium, and rarely in the ventricles.

PRESENTATION OF CASE: We describe a rare case of neonatal cardiac myxoma arising from interatrial septum, causing significant mechanical obstruction to blood flow through tricuspid valve, in an otherwise normal newborn. The patient underwent successful excision of the myxoma with an uneventful recovery [1].

DISCUSSION: Cardiac tumors are rare in children, thus an understanding of the common types of benign and malignant paediatric cardiac tumors and their imaging features, is important because the epidemiology and tumor types differ from those encountered in adults.

CONCLUSION: Large neonatal myxoma is exceptionally rare and even more infrequent is the surgical excision in the first day of life.

1. Introduction

Primary cardiac tumours are rare in paediatric practice, having prevalence between 0.0017 and 0.28 in autopsy series. In contrast, the incidence of cardiac tumours during foetal life has been reported to be approximately 0.14%. The vast majority of primary cardiac tumours in children are benign, whilst approximately 10% are malignant [1,2].

Benign neoplasms are usually classified, according to histologic features and cellular differentiation, as arising from muscle (rhabdomyoma), fibrous (fibroma), vascular (hemangioma), fat (lipoma), nervous (pheochromocytoma), and ectopic tissues (teratoma). However, myxoma and papillary fibroelastoma do not fit into any of the above categories.

Approximately 20% of patients are asymptomatic, with myxomas being found incidentally on imaging of the heart. Clinical features are variable and depend on the location and other associated pathology. However, a triad of symptoms is recognised: valvular obstruction, embolic event and arrhythmias [3].

The clinical presentation is determined by many factors, including tumor location, size, growth rate, tendency for embolization (friability), and degree of invasiveness. Intracavitary tumors tend to obstruct cardiac valves or major vascular structures or produce emboli; myocardial lesions may affect the conduction system of the heart resulting in arrhythmias; pericardial lesions may lead to pericardial tamponade [4].

Echocardiography, Computing Tomography and Magnetic Resonance Imaging of the heart are the main non-invasive diagnostic tools. Tumour biopsy with histological assessment remains the gold standard to confirm the diagnosis.

The differential diagnosis of primary cardiac neoplasia is approached through assessment of the specific location (endocardial/intracavitary, myocardial, or epicardial/pericardial) of the lesion, noting the main chamber or structure involved. The differential diagnosis can be further narrowed by assessing the morphology of the lesion, distinctive radiologic features, and associated imaging findings and correlating them with patient demographics and pertinent clinical information [5]. Secondary involvement of the heart by neoplasia and other non-neoplastic conditions must always be considered and excluded.

Because of the potential life-threatening sequelae of cardiac myxoma, treatment consists of urgent surgical resection. Surgical excision is considered safe, with an excellent long-term prognosis and a low risk of recurrence [6]. It is hypothesized that tumor recurrence is likely related to the development of unsuspected multifocal myxomas rather than an inadequate surgical excision, since only about one-third of resected lesions recur in situ. Because of the risk of recurrence, especially in the group of patients with atypical

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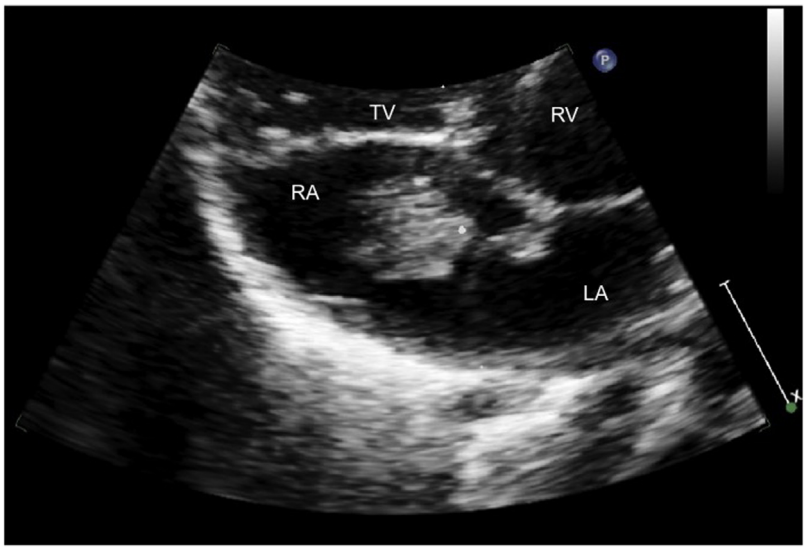

Fig. 1. Right atrial myxoma. Echocardiogram shows a large echogenic mass filling the right atrium.

myxomas, postoperative serial follow-up imaging with echocardiography is usually employed [7].

2. Case report

A newborn in the first day of life was referred to our department with an uncertain diagnosis of a mass floating in the right atrium [Fig. 1]. The patient was known since the 6th month of gestation for a suspected myxoma.

The echocardiography showed an irregular, heterogeneous mass in the right atrium, having a diameter of 12 mm × 10 mm × 8 mm, attached to the interatrial septum, through a 1,5 mm long and thin pedicle, directed to the septum facing the tricuspid annulus [Fig. 2]. The mass was extremely mobile and moved, in diastole, through the tricuspid valve towards the apex of the right ventricle.

Because of the extreme mobility and the thin pedicle of the mass, the baby immediately underwent surgical correction. A normothermic cardiopulmonary bypass was instituted, through median sternotomy, inserting a cannula into the ascending aorta and by use of a double venous cannulae. The right atrium was opened under cardioplegic arrest.

A complete surgical excision of the mass with removal of substantial portion of healthy adjacent endocardial tissue was done.

The mass had the macroscopic appearance of a typical myxoma. It was yellow, soft, polypoid and was attached to the interatrial septum facing the tricuspid annulus in front of the coronary sinus. The mass was completely removed and sent to the pathologist who confirmed it was a myxoma. The patient was discharged on the 8th postoperative day and at 3 months follow up he was in healthy conditions.

3. Discussion

Cardiac tumours may present in foetal or post-natal life. The presenting features depend on the size and location of the mass. In the foetus a tumour can be noted on a routine antenatal anomaly scan as an intracardiac mass. The manifestations of a cardiac tumour in foetal life include arrhythmia, congestive heart failure, hydrops, and not infrequently stillbirth. In postnatal life cardiac tumours may affect the integrity and function of the adjacent cardiac structures leading to severely compromised blood flow due to inflow or outflow tract obstruction, cyanosis, murmur, respiratory distress,

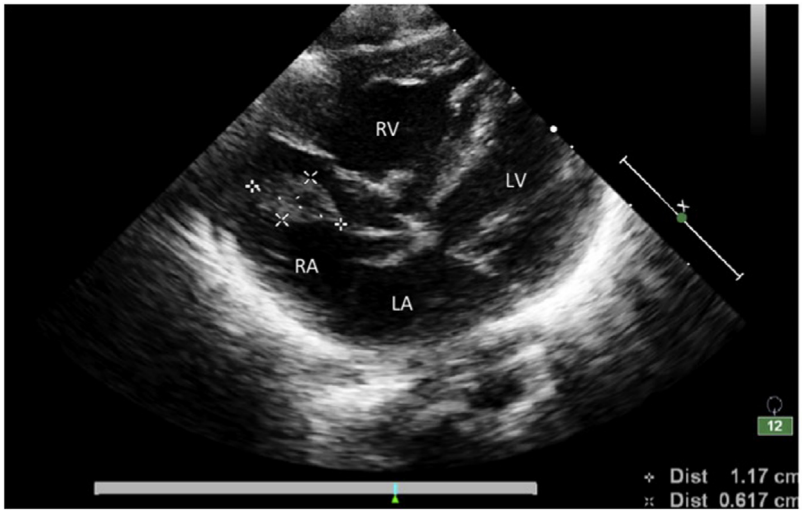

Fig. 2. Right atrial myxoma dimension. Echocardiogram shows a mass attached to the interatrial septum causing flow obstruction.

myocardial dysfunction, valvular insufficiency, arrhythmias, and sudden death.

Due to the progressive nature of pregnancy, fetal cardiac tumours are expected to grow antenatally and it is not unusual for cardiac lesions to be missed at an early obstetric scan. Some tumours can be detected from 20 weeks onwards but the majority will develop later in the course of the pregnancy. Most fetal cardiac tumours will be readily detectable in the late second or third trimester.

Right atrial myxoma has been previously reported in children, which showed symptoms similar to cyanotic heart disease. In rare entity of neonatal cases described in literature, patients expired without having any diagnosis. Often, autopsy finding revealed large atrial mass causing complete obstruction in tricuspid valve area. Cardiac myxoma might cause unspecific general symptoms such as arthralgias, fever, fatigue and anemia, because of released tumor yields such as interleukin-6, tumor necrosis factor or other neuroendocrine factors from the neoplastic tissue. Clinical examinations are otherwise non-specific, however cardiac signs in patients who have no previous cardiac issues in association with general symptoms and embolic events should initiate cardiac investigation. Echocardiography is always considered as first on investigations list. Magnetic resonance imaging generally permits a complete tumor diagnosis and helps planning the operative procedure which is indicated in all patients.

Immediate postoperative mortality ranges from 0.1% to 3.6%. Arrhythmia is a common postoperative complication, which might require long-term medication. Recurrence develops in 2% of the patients and the rate is higher in familial cardiac myxomas.

4. Conclusion

Because primary tumors of the heart in infants and children are extremely rare, most knowledge is based on collections of case reports rather than large cohort studies. The types of heart tumors encountered in the paediatric age group differ from those seen in adults. In the latter, cardiac myxomas are by far the most common tumor.

Despite the fact that myxoma is the most common tumor in adults, it represent an uncommon finding in newborns and the surgical resection in the first hours of life is seldom adopted.

Conflict of interest

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Ethical approval

Ethical approval not need because it was an emergency case.

Consent

Parents of child gave consent for pubblication.

Author contribution

Dr Federica Iezzi, Dr Andrea Quarti: writing the paper.
Dr Alessandro Capestro: data analysis.
Dr Marco Pozzi: revision of the paper.

Guarantor

Dr Federica Iezzi.

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