

Cardiac Surgical Challenges in Management of a One Day Old Infant Presenting with Large Right Atrial Tumour

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Abstract

Though it is established that certain primary tumours of the heart may be present in infancy, little information is available recommending best practice for various surgical issues that may present during the first week of life. We report a very unusual intra-cardiac tumour presentation with challenges both clinically and pathologically.

Keywords: Atrial tumour; Myxoma; Rhabdomyoma

Introduction

Although certain primary tumours of the heart may be present in infancy, there is little data accessible regarding suitable practical recommendations for the various surgical challenges that may present during the first week of life. We report an unusual intra-cardiac tumour presentation from a clinical and pathological perspective: its position, size, clinical impact, and the surgery in an emergent setting were uniquely challenging. Confirmation of tumour type by features, typical appearance, or by frozen section histological facilities (unavailable) was not possible. Rhabdomyomas are not well characterized but are usually multi-centric. Myxomas do not usually arise from the lateral wall of the right atrium. This tumour was sessile, uni-centric, and had an extensive broad attachment to the lateral right atrial wall, rising into the territory of the sino-atrial node. Emergent surgery took place in a clinical cardiac program that lacks advanced cardiac investigative and surgical facilities [1].

Case Study

A one-day old female patient, with normal appearance, no familial history nor features suggestive of a genetic abnormality, was the second child born in a rural location in Ecuador. Gestational progress was normal, and the mother denied alcohol, drugs or smoking during pregnancy but, lives on a farm with known pesticide exposure. At birth, following uncomplicated vaginal delivery with an APGAR score of 10, weighing 3 kg, the neonate was found to have sustained cyanosis, edema, and congestive cardiac failure, within 24 hours; ventilatory and inotropic support was established for the infant with no improvement which necessitated the decision to proceed with early surgical intervention. Two-dimensional echocardiography showed a 2 × 2.5 cm right atrial tumour arising from the right lateral wall and sub-total blockage of the superior and inferior venae cavae (SVC, IVC), and tricuspid valve. No other chamber nor the myocardium were similarly affected (Figure 1). There were substantial pericardial, pleural and peritoneal effusions with hepato-splenomegaly. Hematological profile was unremarkable, and no rhesus incompatibility was identified [2].

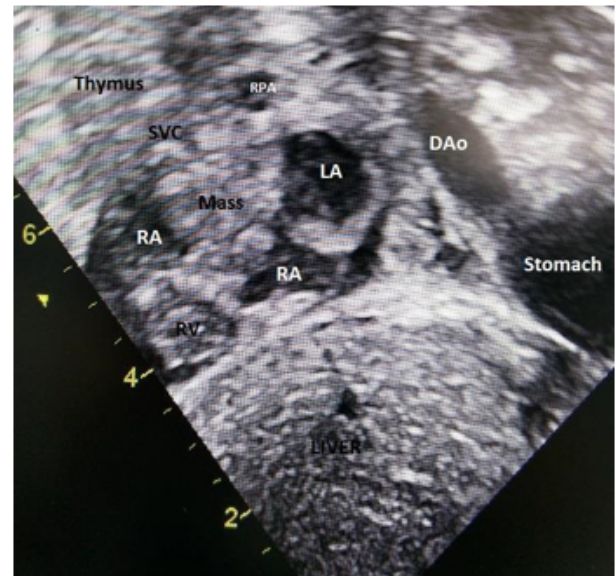


Figure 1: Two-dimensional echocardiogram, showing the mass in the right atrium.

Following pleural drainage yielding straw-coloured fluid, hypothermic cardiopulmonary bypass was instituted with difficulty; it required high SVC cannulation to initiate. Upon SVC snaring, the right atrium was opened, and sucker drainage of the IVC was later accomplished by careful retraction of the mass. The tumour had a variegated, polymorphic appearance and was tightly attached to the lateral wall; no separation plane could be established. It was removed with 2 × 2.5 cm of the lateral wall, sacrificing half the sino-atrial area at the SVC/atrial junction. Autologous pericardium was sutured in to close the defect and provide sufficient atrial cavity size [3].

Post-operatively, limited evaluation of neurological status appeared normal, and hemodynamic recovery was accomplished. Even though

an extensive resection was made, compromising half of the SA node, sinus rhythm returned after several hours and signs of congestive cardiac failure diminished during the early post-operative period. The patient recovered in the ICU with the chest open until post-operative day-5 (POD). On POD 7 patient became vasoplegic due to severe sepsis, refractory to inotropes and vasopressors resulting on the patients demise on POD-10.

Pathological evaluation after staining and a second pathology expert opinion, confirmed the tumour was a rhabdomyoma (Figure 2). Although formal examination of the surface of the heart, and right ventricular cavity had revealed no other lesion (rhabdomyomas are usually multi-centric), this presentation was very unusual. At surgery, the position and sessile attachment of the tumour suggested myxoma, but myxomas have not been reported arising from the lateral wall.

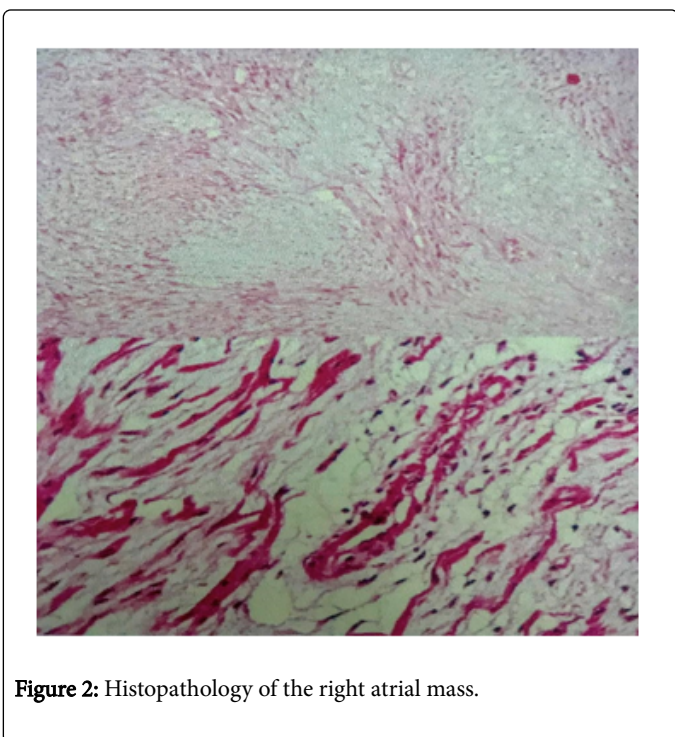


Figure 2: Histopathology of the right atrial mass.

Discussion

Cardiac tumours are abnormal growths in the heart chambers or in the valves. They may present as, primary (Benign or malignant), or

metastatic (Malignant). Myxomas are the most common primary heart tumour overall; they are benign in nature, frequently found in the left atrium at the level of the inter-atrial septum and can also originate from the anterior or posterior wall, or the left atrial appendage; a smaller percentage arise from the right atrium. They usually present as a solitary, round or oval, polypoid mass; they can cause mechanical obstruction, embolization, arrhythmia and pericardial disorders.

Rhabdomyomas are a type of striated muscle tumour, hamartomatous in origin, and the most common cardiac tumour in children. They may present as multiple lesions, are intra-mural or intra-cavity, and are predominantly located in the ventricles. They can cause chamber obstruction or arrhythmias. They are strongly associated with Tuberous Sclerosis. The diagnosis of cardiac masses can be made primarily by echocardiography. Other types of cardiac tumours seen include: fibromas, teratomas, hemangiomas and cardiac fibromas.

Conclusion

Emergent surgery is necessary and can be accomplished in such clinical presentations despite lacking ideal operative facilities and resources. Adjustment of standard surgical bypass procedures to deal with the physical and functional blockage to blood flow is required. Surgical excision without the benefit of tumour identification may result in diseased tissue being left behind. Sino-atrial nerve dysfunction and intra-cardiac edema may complicate post- surgery management as congestive cardiac failure resolves. In this presentation, sepsis was the main contributing factor that led to the demise of the patient.

Conflicts of Interest

There are no conflicts of interest for the present study.

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