Right atrial aneurysm treated with atrioplasty without using cardiopulmonary bypass in an infant

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Abstract

A 7-month-old female child was diagnosed to have a massive right atrial aneurysm, presented with arrhythmias. She was treated by restorative right atrioplasty without using cardiopulmonary bypass. She had good postoperative outcome and a 5-month follow-up remains uneventful. In the absence of major congenital heart defects, aneurysm of right atrium can be treated without using cardiopulmonary bypass.

Keywords: Aneurysm; Atrioplasty; Right atrium

1. Introduction

Aneurysms of the right atrium may be congenital or acquired. It usually presents with arrhythmias or as an additional finding in association with other congenital lesions in the heart. Due to rarity of its presentation, the optimal line of management has been unclear.

2. Case report

This 7-month-old girl who weighed 3.4 kg was referred for recurrent supraventricular tachycardia, mainly atrial flutter with regular ventricular conduction. Antenatal diagnosis of dilatation of right atrium of uncertain aetiology was already made in this case. The delivery of the child was uneventful. On follow-up, she was found to have tachyarrhythmia for which she was treated with Beta-blockers and aspirin.

On examination, physical findings were unremarkable. X-ray chest showed severe cardiomegaly. Electrocardiogram depicted runs of supraventricular tachycardia. Echocardiography revealed massively dilated right atrium with a circumference of 21.6 cm and an area of 29.6 cm² and a small patent foramen ovale with left to right shunt. No signs suggestive of Ebstein anomaly were found. Left ventricle function was normal.

Operation was performed by median sternotomy. At operation, the findings were remarkable. Right atrium was massively dilated, occupying most of the pericardium and completely obscured the view of aorta, pulmonary artery and right ventricle (Fig. 1). Right atrial wall was thin, floppy and akinetic. Restorative right atrioplasty was performed by progressively excising the aneurysm wall from four different places, utilizing a soft vascular clamp and suture closure of atriotomy with running 6-0 Prolene stitch (Fig. 1). The incision line on atrium was designed to simulate the atrial wall incision of right-sided Maze procedure. However, no cuts were performed on atrial septum as cardiopulmonary bypass was not used. Right atrial appendage was preserved.

She recovered very well postoperatively and postoperative X-ray showed significantly reduced cardiac size (Fig. 2). Postoperative echo suggested right atrial size of 8–10 cm² and trivial tricuspid regurgitation. She remained in sinus rhythm after the operation.

Histology of excised right atrial wall showed extensive fibrosis with myxoid appearance. Vacuolations in the myocardial bundle were suggestive of myocytolysis. Vacuolations seen in this case were similar to what has been seen with histiocytoid cardiomyopathy.

3. Discussion

Aneurysm of right atrium is a quite uncommon condition, which has been reported as cases detected in age groups varying from neonates to late adulthood [1]. It is identified under various headings like aneurysm, diverticulum or idiopathic dilatation. Baily et al. were the first to report the condition [2].
Majority of patients present with atrial arrhythmias and symptoms associated with it. Associated congenital heart defects consisting of atrial septal defect, ventricular septal defect and coronary sinus diverticula are not uncommon [3,4]. It is obvious that there is a congenital structural defect in atrial wall which makes it prone to dilations even with right-sided pressures which are lower compared to the left side. Lipomatous degeneration and reduction of muscular elements in the aneurysmal wall have been reported [1]. Histiocytoid cardiomyopathy is reported as one of the rare conditions causing tachyarrhythmia in children [5].

Literature reports various ways to manage this condition like observational management, resection using cardiopulmonary bypass and correction of associated defects. Conservative management may not prevent atrial dilatation which could invite complications of thromboembolism, fatal arrhythmias, tricuspid regurgitation and sudden death. Arrhythmias have been successfully treated by surgical excision of aneurysm [6-8]. Kopf et al. used Maze procedure quite effectively for complex right-sided congenital heart diseases and concluded that Maze operation is one of the effective way to control arrhythmias, especially re-entry tachycardia in children who are going to have operation for associated congenital heart defect, and also suggested for further investigation due to shorter follow-up in their study [9]. This study and other similar studies are yet to address the issues with type of incisions, biatrial versus right atrial procedure and selecting a type of an energy source. Decision-making is easy when the patients have other congenital defects which mandate surgical intervention. This patient had isolated right atrial aneurysm without any major congenital defect. Maze procedure is probably more suitable for patient with right-sided lesions like Ebstein anomaly, tricuspid atresia and re-operation for lateral tunnel Fontan procedure. Addition of Maze procedure in our patient would complicate the relatively simple procedure, which could be done without using bypass machine. Her enormously dilated right atrium was causing clinically important recurrent atrial flutter with tachycardia requiring medical treatment.

She underwent operation without the use of cardiopulmonary bypass as she did not have any major congenital heart defects. In the absence of associated defects, it is easy to resect the aneurysm by applying soft vascular clamps of different shapes, and systemic heparinization is not necessary. The recovery is faster and the complications related to cardiopulmonary bypass can be easily avoided. We also preserved her right atrial appendage which may be useful in preservation of atrial natriuretic factor (ANF) and secure cannulation site for future operations if the need arises. In this patient, histology was suggestive of myocytolysis with few muscle fibers in the excised atrial wall which would make right atrium relatively less contractile making it prone to progressive dilatation.

We believe that atrial arrhythmias in these cases are mostly secondary to atrial dilatations and structural disorientation of fibers and probably conduction system, particularly in cases where there has been no definite histopathology found. Excision of parts of atrial wall involved in aneurysm would interrupt the re-entry pathways and in someway helps in controlling re-entry arrhythmias. We recommend early treatment of clinically significant aneurysms of right atrium, especially in a child associated with atrial arrhythmias. Atrial appendage should be preserved for reasons stated above and surgery can be done without using cardiopulmonary bypass if patient does not have major congenital heart lesions. Avoidance of cardiopulmonary bypass should reduce the postoperative morbidity which is difficult to quantify due to lack of randomized study in infants. Progressive dilation and recurrence of arrhythmias remain a possibility which necessitates long-term follow-up of these patients.

References


