Case Report

Giant Right Atrium - A Rare Case Report
Nanda Kishore, MD, Ravi, MD, Nagaraj Gowda, MD, Devananda, Mch
Departments of Cardiothoracic Anaesthesiology, and Cardiothoracic Surgery,
Manipal Heart Foundation, Bangalore

Various types of congenital malformations of the right atrium (RA) and coronary sinus have been described. They can be congenital enlargement of the RA, single or multiple diverticulae of RA and diverticulae of coronary sinus. They can be asymptomatic and incidentally detected on a chest radiograph with large cardiac silhouette. We describe a child who presented with symptomatic giant enlargement of the RA.

Case Report

An 8 month old child weighing 7 kg was referred from the paediatrics department with respiratory failure. This was a term baby, delivered by caesarean section because of oligohydromnios. Foetal echocardiography during the antenatal period had revealed RA dilation. APGAR score at birth was 8/10 at 1 min and 9/10 at 5 min. The child had several episodes of respiratory distress requiring repeated hospitalization and even temporary ventilatory support during the course of medical management. Flexible bronchoscopy did not reveal any pathology attributable to the symptoms of child. Serial echocardiography performed during this period showed gradual increase in the size of RA. Since the size of RA was increasing and the child was symptomatic he was referred for surgery. Physical examination showed respiratory rate of 60/min with subcostal and intercostal indrawing and bilateral pulmonary rhonci. Heart rate was 180/min with normal jugular venous pressure. An ejection systolic murmur was heard at pulmonary area. On abdominal examination, there was no organomegaly. Arterial oxygen saturation (SpO₂) was 98% with 4 liters/min of oxygen by mask.

Chest X-ray showed gross cardiomegaly (Fig. 1). Preoperative echocardiography (Fig. 2) revealed giant RA, ostium secundum atrial septal defect (ASD) with bi-directional shunt, dilated right ventricle (RV) and moderate pulmonary arterial hypertension (PAH). Other laboratory investigations were within normal limits. Child was receiving lacciactone, digoxin and antibiotics and was scheduled for open-heart surgery for reduction atrioplasty and ASD closure on an urgent basis. No preanaesthetic medication was given. On arrival in the operation theatre, monitoring with ECG, SpO₂, noninvasive blood pressure, and peripheral skin temperature was started. Oxygenation by mask was continued.
Anaesthesia was induced with intravenous fentanyl citrate 20 µg, midazolam 0.25 mg and endotracheal intubation was accomplished with vecuronium 1 mg. Anaesthesia was maintained with 100% oxygen and titrated doses of fentanyl, midazolam, vecuronium and isoflurane. Femoral artery and vein were cannulated for arterial pressure monitoring and central venous access.

After institution of routine hypothermic cardiopulmonary bypass, cardiac anatomy was reassessed. Surgical findings included giant thin walled right atrium and large ostium secundum ASD. ASD was closed with pericardial patch and reduction atrioplasty was performed. Total cardiopulmonary bypass time was 65 min and aortic cross clamp time 22 min. Patient was weaned off cardiopulmonary bypass with minimal inotropic support and extubated in the early postoperative period. Postoperative chest X-ray and echocardiographic pictures are shown in figure 3 and 4 respectively. Further postoperative course was uneventful.

Discussion

Congenital malformations of RA and coronary sinus are rare, and only sporadic cases have been reported. Little is known about the clinical relevance of this disorder. The present case had gross enlargement of the RA with ostium secundum ASD which was detected by foetal echocardiography. Follow-up echocardiography revealed progressive increase in the size of RA.

Clinical presentations vary depending on the type of RA malformation. Those with enlargement of RA and large diverticula frequently come to medical attention because of gross cardiomegaly on radiograph. They can also present with atrial arrhythmias. Patients with diverticulae of coronary sinus usually present with arrhythmias associated with accessory pathways that traverse the diverticulum to form an atrioventricular connection. Sudden deaths also have been reported. The present patient had symptoms related to congestive cardiac failure and needed repeated hospitalization.

Surgical reduction atrioplasty is required in most cases. Those with significant tricuspid regurgitation require tricuspid annuloplasty. As the child was symptomatic, surgical intervention was contemplated. Treatment of asymptomatic patients is controversial. Since they are at high risk of developing thrombus and atrial arrhythmia, anticoagulation is necessary. In patients with arrhythmias, surgical ablation of accessory pathway may be required. The enlarged RA is described as being “paper thin” and translucent in several patients. Demarco and Bolero have described the largest atria with volume of 900 cm³. This pathologic appearance bears a striking resemblance to the description of cases of so called “Parchment Heart” or Uhl’s anomaly. In Uhl’s anomaly there is marked myocardial hypoplasia or aplasia involving one or more cardiac chambers.

The wall of enlarged RA and RA diverticula
often demonstrate lipomatous degeneration and reduction of muscular elements. In the present case microscopic examination of the sections from right atrial wall showed myocardial tissue as well as fibro-collagenous tissue on its exterior surface. Myocardial cells showed mild oedema. Enlarged RA can cause symptoms related to compression of intrapericardial structures such as jugular engorgement, oedema and hepatomegaly. This may influence the metabolism of anaesthetic drugs and choice of drugs and dosage should be made judiciously. The use of fentanyl, midazolam and vecuronium appears to be appropriate and was used in the present case.

The enlarged RA can get injured during sternotomy and the whole operating team should be ready to deal with such a complication. The anaesthetist should be ready to transfuse large volumes and the surgeon and the perfusionist ready to establish bypass urgently.

In conclusion, a symptomatic child with giant RA and ostium secundum ASD undergoing surgical repair was successfully managed. In addition to the usual precaution of conducting bypass in an infant, judicious use of fentanyl, midazolam and vecuronium appears to be safe in such a patient.

References

4. Beder SD, Nihill MR, McNamara DG. Idiopathic dilation of the right atrium in a child. Am Heart J 1982; 103: 134-137