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SURGICAL PATCH CLOSURE OF RIGHT PULMONARY ARTERY TO LEFT ATRIUM FISTULA - A RARE ENTITY WITH COMMON CLINICAL PRESENTATION.

Dr. Lata Khatnani Koneru* Mch Cardiothoracic Surgery, Department of CTVS, Dr. Ram Manohar Lohia Institute of Medical Sciences, Lucknow, India. *Corresponding Author

Dr. Anurag Rai Department of CTVS, Dr. Ram Manohar Lohia Institute of Medical Sciences, Lucknow, India.

Dr. Subhash S. Rajput Professor & Head of the Department of CTVS, Dr. Ram Manohar Lohia Institute of Medical Sciences, Lucknow, India

ABSTRACT Direct fistulous communication of the right pulmonary artery and the left atrium is a rare congenital malformation of the pulmonary vasculature. It presents as either central cyanosis or its complications and heart failure specially in newborn. Intervention (whether transcatheter or surgical) is usually determined by the anatomy, location and type of the fistula.

This report illustrates a case of an eight year old male child presented with cyanosis and dyspnoea, diagnosed to have Right Pulmonary Artery to Left Atrium Fistula with large Atrial Septal Defect on various imaging studies. Location of fistula opening was near left pulmonary veins. In this scenario Transcatheter closure can cause pulmonary vein obstruction hence Surgical repair was opted and done successfully.

KEYWORDS : central cyanosis, arteriovenous malformation, right pulmonary artery to left atrium fistula.

Introduction

Right Pulmonary Artery to The Left Atrium Fistula [RPALAF] is a rare congenital anomaly [1]. True incidence is unknown [2]. Clinically patients present initially with cyanosis, later in untreated cases with chronic hypoxemia, brain abscess, systemic thromboembolism, or cerebrovascular accidents [1]. Diagnosis of this entity needs careful clinical examination, expert echocardiography and Cardiac angiographic evaluation. Various surgical methods of management of these cases have been described in the literature depending upon its type [2]. These cases can be managed surgically with either transthoracic approach without CPB or median sternotomy approach with or without CPB. Recently transcatheter closure with a device, coils or with vascular plug has also been described [3,4,5].

In our case, orifice of fistula at LA side was closed internally and reinforced with PTFE patch successfully.

Case report

An eight year old male child referred for central cyanosis and dyspnea since 3 year of his age. Clinical examination revealed, central cyanosis, clubbing, failure to thrive with BSA 0.85 m2 (Du Bois formula). On auscultation patient had wide and fixed split S2, and grade III ejection systolic murmur at pulmonary area. His spo2 was 65%, hemoglobin and hematocrit was 23.4gm/dl and 74% respectively. Chest X-Ray showed cardiomegaly with cardiothoracic ratio of 0.6 and crescent shape opacity parallel and outside RA shadow. ECG showed normal sinus rhythm with a heart rate of 95 beats/mins, Bi-atrial enlargement with RV volume overload, incomplete RBBB. All other laboratory findings were within normal limits.

Transthoracic Echo study showed situs solitus, AV-VA concordance, large Ostium Secundum Atrial Septal Defect (OS ASD). Normal systemic venous drainage, abnormal right pulmonary artery drainage to LA through fistulous communication, dilated RA/ RV & aneurysmally dilated LA [Fig 1].

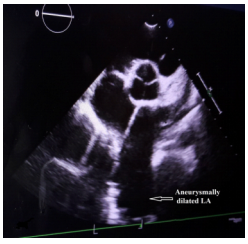

Figure 1 – Transthoracic echocardiographic image in short axis parasternal view showing aneurysmally dilated LA.

Bubble contrast study revealed dense opacification of the left atrium within 2 cardiac cycles suggests direct communication with the venous circulation.

Cardiac CT angiography was done to delineate detailed cardiac anatomy.

It showed RPA to LA fistula opening near the roof of LA close to upper pulmonary veins with aneurysmal dilatation of Left atrium, normal pulmonary venous drainage [Fig. 2a, 2b].

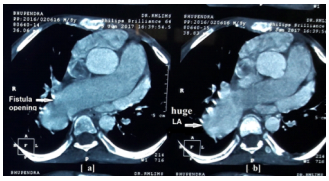

Figure 2: (a and b) CT pulmonary angiographic images in horizontal plane showing RPA to LA fistula origin (arrow) and a wide direct communication between RPA and the left atrium with aneurysmally dilated huge LA.

Transcatheter Device closure was excluded in the view of the possibility of obstruction of the pulmonary veins. Surgical closure was planned.

Operative steps-

Conventional median sternotomy was done with standard ascending aortic and bi-caval cannulation. Right pleura opened and examined, part of dilated LA was embedded in right lung hilum and parenchyma. Fistulous communication could not be identified externally as dense adhesions were present. Hence decided to close it internally through LA. For clear surgical field, vent (sump catheter) was inserted in MPA and then aorta was cross clamped. Heart was arrested with hyperkalemic hypothermic blood cardioplegia. RA opened after arresting the heart. Intracardiac anatomy inspected. Near the junction of LA roof and LSPV opening, sump catheter tip was seen through fistulous opening (Fig 3).

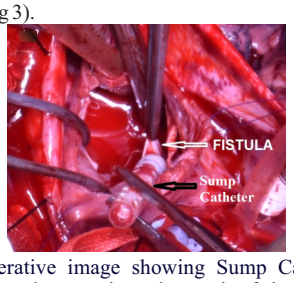

Figure 3 - Intraoperative image showing Sump Catheter (MPA vent)(black arrow) coming out through mouth of the fistula (white arrow) confirming site of fistula and direct communication.

It confirmed the site of communication. Opening was closed with 5-0 polypropylene purse string suture reinforced with PTFE patch. Patch was sewn with 5-0 polypropylene sutures in continuous manner. ASD patch (PTFE) closure and RA closure was done in usual manner. CPB support was weaned off slowly. Post CPB there was increase of saturation (SPO2) from 67% to 100% and PaO2 increment was from 72 to 250 at 0.5 FIO 2 (fractional inspiratory oxygen).

Discussion

Fistulous communication between the right pulmonary artery (RPA) and the left atrium (LA) was first reported by Friedlich et al in 1950 [6]. He observed that due to the low resistance in (anomalous) channel, blood flow was from RPA to LA leading to cyanosis with increased pulmonary blood flow. Since than fewer than 100 cases have been reported [7]. Embryologically, it develops due to incomplete degeneration of the septum between the arterial and venous plexus of the pulmonary vascular bed. A pulmonary vein connected to such a fistula is absorbed into the LA during development, forming a PA–LA fistula [2]. In most of the reported cases, the anomalous vessel originates from the right pulmonary artery after its middle branch and is connected to the left atrium between the right and left pulmonary veins. In our case anomalous vessel was arising from distal part of right pulmonary artery and was opening near left superior pulmonary vein.

De Souza e Silva [8] classified it in three types, depending upon pulmonary venous drainage pattern. Fourth type was later described by Ohara et al [9].

Type I: Normal pulmonary venous drainage pattern.
Type II: Absent right inferior pulmonary vein with fistulous connection at the normal site of its origin. associated with right lung abnormalities.
Type III: All pulmonary veins connected to the aneurysmal pouch.
Type IV: Right inferior pulmonary vein replaced by three small veins connected to the aneurysmal pouch.

In our case, CT angiography revealed it Type I fistulous communication.

Associated atrial septal defects are common. Other concomitant cardiac malformations are rare. Clinical diagnosis of this anomaly is difficult. The age of presentation ranges from 1 month to 60 years depending upon the size of fistula and magnitude of shunt. Bubble Contrast echocardiography reveals contrast in the left atrium within 2-3 cardiac cycle suggests direct communication with the venous circulation. Differential diagnosis which should be considered with similar clinical and echocardiographic scenario are, a left superior vena cava that drains into the left atrium, an unroofed coronary sinus, or an atrial septal defect with elevated right atrial pressure and a right-to-left shunt.

In our case, echocardiographic evaluation showed a right-to-left shunt at LA and RPA, an atrial left-to-right shunt at ASD (interatrial) and no sign of elevated pulmonary pressure. Echo was inconclusive about cardiac fistulous anatomy; hence Cardiac CT was necessary to confirm this diagnosis.

Transcatheter Device closure was excluded in the view of the possibility of obstruction of the pulmonary veins as orifice of fistula was close to Left Superior Pulmonary Vein.

From a surgical point of view, the lesions have been approached through median sternotomy, right posterolateral thoracotomy, or left posterolateral thoracotomy depending upon type and site of fistula [2]. Through median sternotomy extracardiac simple ligation or division and intracardiac closure both are possible. In our case we approached through median sternotomy. External ligation or division was technically difficult hence intracardiac approach was decided. The site of fistula in LA, accidently guided by MPA vent, due to different magnitude of resistance allowed passage of vent through the low resistance (anomalous) channel. Sump catheter tip was seen at orifice of fistula, which was the confirmatory guide to fistula. After confirmation Successful intracardiac closure of communication was done.

Conclusion

We conclude that in evaluating patients of central cyanosis we should have high level of suspicion of this rare anomaly as early diagnosis and intervention can decrease morbidity and mortality associated with this anomaly. Peculiar Chest X ray and echo findings give a clue, regarding diagnosis of such a rare anomaly. However for correct diagnosis and evaluation of cardiac anatomy, cardiac CT angiography is mandatory in present era.

In our case diagnosis was made preoperatively, based on echo n CT angiogram findings and preplanned for surgical closure. However insertion of MPA vent guided accidentally and it showed orifice of fistula. In future MPA vent insertion can be used for determining the site of communication in difficult and complex cases.

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