A rare cause of Right ventricular outflow tract Pseudoaneurysm in Tetralogy of Fallot

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Abstract

Pseudoaneurysm of the right ventricular outflow tract(RVOT) is an uncommon yet catastrophic complication after intracardiac repair of Tetralogy of Fallot(TOF). We describe a patient diagnosed with RVOT pseudoaneurysm in the immediate postoperative period after complete repair for TOF with single pulmonary artery. The pseudoaneurysm was repaired successfully. This case is reported to emphasise the importance of a high degree of suspicion of this rare entity in these patients for its early diagnosis and management.

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Abstract

Pseudoaneurysm of the right ventricular outflow tract(RVOT) is an uncommon yet catastrophic complication after intracardiac repair of Tetralogy of Fallot(TOF). We describe a patient diagnosed with RVOT pseudoaneurysm in the immediate postoperative period after complete repair for TOF with single pulmonary artery. The pseudoaneurysm was repaired successfully. This case is reported to emphasise the importance of a high degree of suspicion of this rare entity in these patients for its early diagnosis and management.

Key words: Pseudoaneurysm, Tetralogy of Fallot, Single pulmonary artery, Surgical correction

Case report:

A 3 year old male child was admitted with complaints of recurrent cyanosis for the past six months. He was diagnosed with Tetralogy of Fallot(TOF) and planned for surgical repair. Echocardiography and cardiac catheterisation confirmed the diagnosis of Situs solitus, Levocardia, TOF with absent left pulmonary artery and severe infundibular stenosis. Left lung being supplied by smaller Aortopulmonary collaterals. His pre-operative hemogram, renal function and liver function were within normal limits.

After obtaining parental consent, child was taken up for surgical correction. Bi-ventricular repair was done that include Dacron patch closure of ventricular septal defect, resection of hypertrophied septal and parietal bands, reconstruction of right ventricular outflow tract(RVOT) with autologous pericardial patch. The pericardium was treated using 0.6 % glutaraldehyde for eight minutes and was fixed using 6-0 prolene single layer full thickness suture. Pulmonary annulus was accepting Hegars no.11(Z score -2) thereby the native tricuspid pulmonary valve was preserved. Post repair, pressure ratio of Right ventricle to left ventricle was 0.6 and gradient across pulmonary valve was 32 mmHg. Child underwent tracheal extubation on day 2, mediastinal drains were removed on day 3 and remained hemodynamically stable.

Child remained asymptomatic but started developing ill-defined opacity in left hemithorax on x-ray from the fifth postoperative day(figure 1). Since it was persisting even after 48 hours and was gradually increasing in size, Contrast Enhanced Computed Tomography(CECT) was done that described a large Pseudoaneurysm of size 55x35 mm arising from RVOT, occupying almost two thirds of left pleural cavity(figure 2). After explaining the risks, child was shifted for Pseudoaneurysm repair. Right Femoral vessels were exposed

and redo sternotomy was done, Cardiopulmonary bypass(CPB) was initiated with aortic and right atrial cannulation. After moderate hypothermic cardioplegic arrest, RVOT patch was found to be completely disrupted and blood was seeping into the aneurysmal cavity. The previous patch and the aneurysmal tissue(figure 3) were completely excised, edges freshened up and was reconstructed using bovine pericardium in two layers. The patient was weaned off CPB with minimal inotropic support and was on mechanical ventilatory support for 48 hours.

Child underwent extubation on day 3 and had an uneventful postoperative course thereafter. The occurrence of RVOT Pseudoaneurysm in a postoperative patient of TOF with absent left pulmonary artery is due to its atypical morphology and has not been reported in literature so far.

Discussion:

Pseudoaneurysm of the RVOT is an uncommon postoperative complication of pediatric cardiac surgeries that involve both a right ventriculotomy and RVOT reconstruction with patch repair or conduit replacement [1-3]. The various mechanisms of pseudoaneurysm formation has been described as related either to elevated residual right ventricular systolic pressure or obstructive RVOT, leading to mechanical strain at the proximal suture line and the use of patches, homografts, or conduits to reconstruct the outflow tract, with dehiscence of surgical sutures being the aetiology of the pseudoaneurysm. However, other factors such as suturing technique, trauma, complete heart block, and infection have also been rarely implicated[1-3]

Regardless of the aetiology, a pseudoaneurysm arises from the small dehiscence of a portion of the reconstructed RVOT, permitting blood seepage into the surrounding pericardial or pleural cavity [3,4]. In the index case, after excluding infection, heart block, and surgical technique, the pseudoaneurysm might have been caused by the exceptional morphology that is, absent left pulmonary artery with unusually high position and acute angulation of pulmonary valve with RVOT(figure 4) supplemented partly by RV hypertension.

Pseudoaneurysms usually remain asymptomatic and are found incidentally on follow-up[3]. However, they may present rarely with secondary symptoms due to compression of adjacent mediastinal structures, thromboembolism and infection [3,4]. In the present case, it was an incidental finding on chest x-ray. Though transthoracic echocardiography is a reliable method for diagnosing pseudoaneurysms[5] it does not yield definitive information regarding the extent and relationship of it with the surrounding structures. CECT and Magnetic resonance imaging [6] are more precise in furnishing the size, extent and location of the pseudoaneurysm, its anatomical relationships and retrosternal anatomy. Once diagnosed, surgery is the treatment of choice.

Conclusion:

A pseudoaneurysm in a surgically reconstructed RVOT is rare. It should always be suspected in patients with unusual morphology such as TOF with Single pulmonary artery and those with RV hypertension and such patients must be followed up with high suspicion even in the immediate postoperative period and long term.

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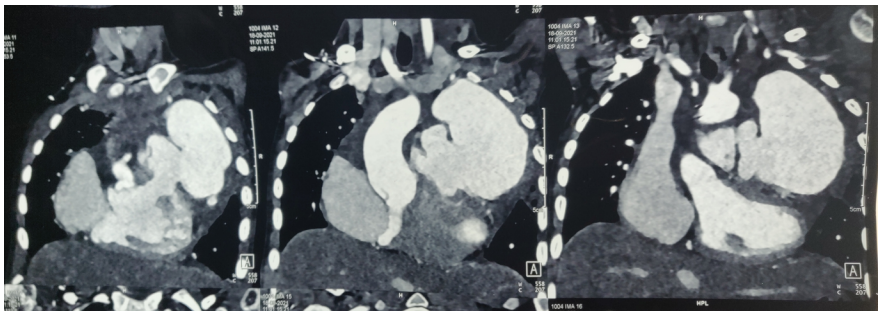
Figure legends

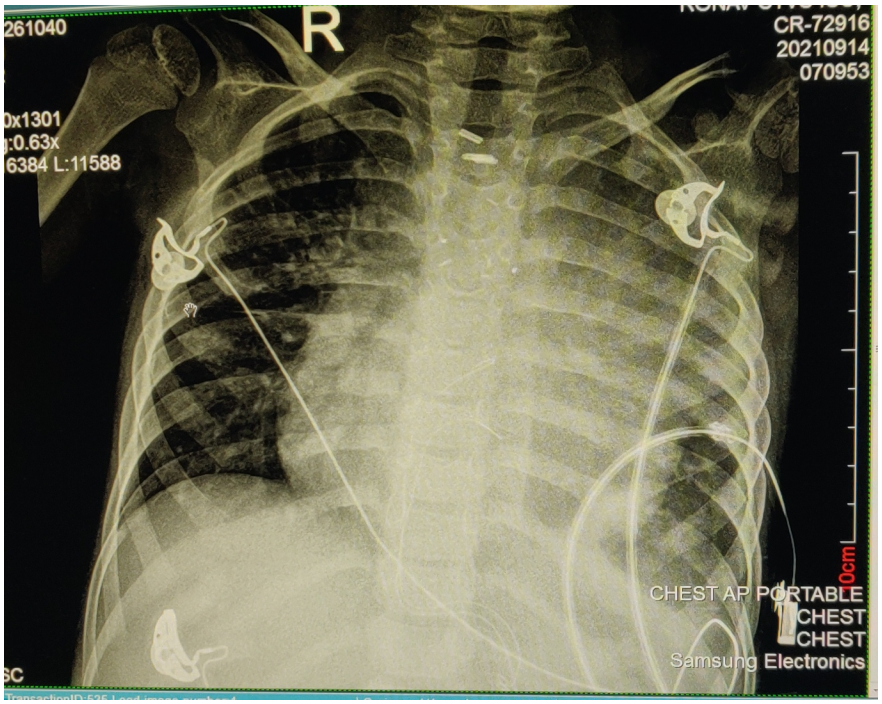
Figure 1: Chest x-ray AP view showing opacity in the left hemithorax

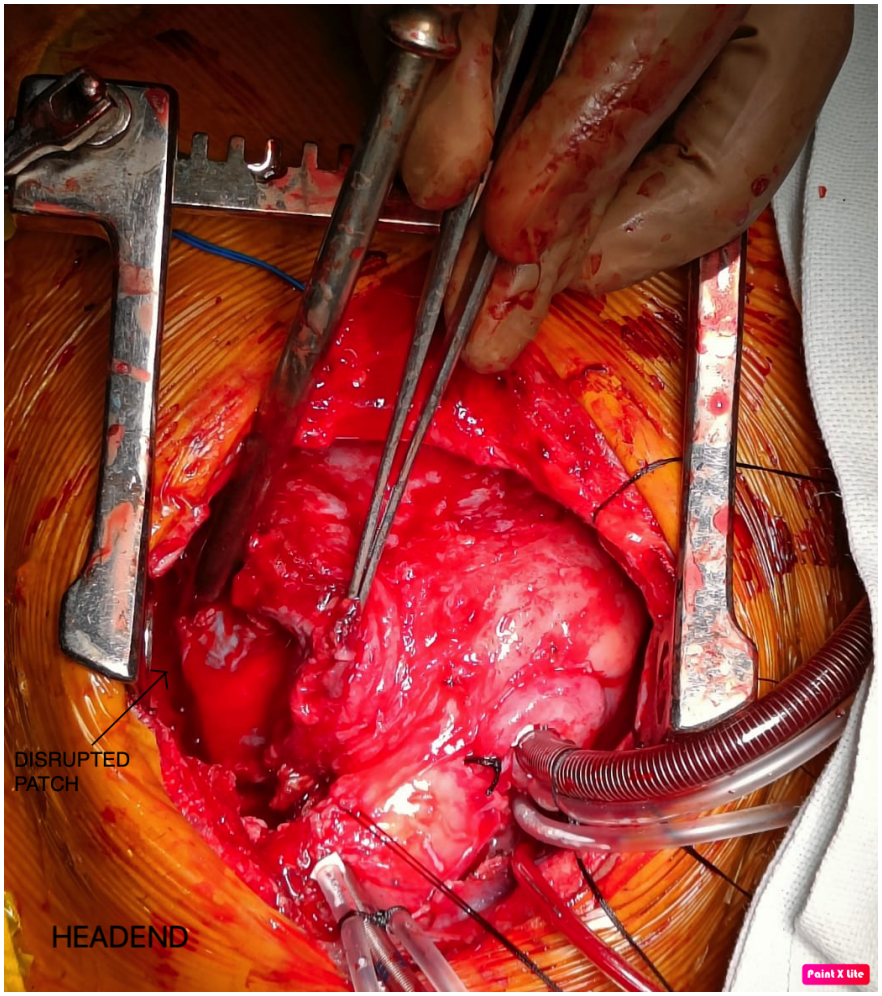
Figure 2: Coronal sections of cardiac CT showing a large Pseudoaneurysm arising from RVOT

Figure 3: Intra-operative image showing disrupted RVOT patch

Figure 4: Transesophageal echocardiography aortic short axis view showing relatively the higher position of the pulmonary valve with its continuation into the right pulmonary artery







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