Right ventricular outflow tract aneurysm with thrombus

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Abstract

Right ventricular outflow tract (RVOT) aneurysm is a known complication of tetralogy of Fallot repair when a ventriculotomy is done. It leads to RV dysfunction and may require re-operation. We describe a rare instance of a patient who developed an RVOT aneurysm after trans-ventricular repair of tetralogy of Fallot, which was complicated with the formation of a thrombus in the aneurysm sac. The patient underwent re-operation with thrombectomy, excision of the RVOT aneurysm and pulmonary valve replacement. To the best of our knowledge, the occurrence of this combination and its implications have not been reported.

Keywords: Congenital Heart Disease • Tetralogy of Fallot • Aneurysm • Pulmonary valve

INTRODUCTION

Surgical correction of tetralogy of Fallot has excellent long-term survival [1]. Long-term complications of the right ventricular outflow tract (RVOT) are the main cause of re-operation [2]. RVOT aneurysms are seen in about 6–25% of the patients who undergo tetralogy of Fallot repair [3].

Here, we report a rare case of a postoperative RVOT aneurysm, which was complicated with thrombus formation in the aneurysm sac. To the best of our knowledge, this is the first report in English literature.

CLINICAL SUMMARY

A 25-year old female who had undergone trans-ventricular tetralogy of Fallot correction with reconstruction of the RVOT with untreated pericardial patch 3 years back presented to us with New York Heart Association class III breathlessness and easy fatigability of 3 months of duration. On physical examination, the patient was afebrile and haemodynamically stable with a room air oxygen saturation value of 96%.

Chest X-ray showed a cardiothoracic ratio of 60% with prominent shadow over the left heart border inferior to the aortic knuckle (Fig. 1a). Echocardiography showed severe RVOT obstruction with a mean gradient of 86 mmHg, aneurysmal dilatation of the RVOT, moderate tricuspid regurgitation and RV dysfunction. The pulmonary valve leaflets were thickened, and there was no pulmonary regurgitation. To ascertain the level of RVOT obstruction, a cardiac catheterization was performed, which confirmed the diagnosis of an RVOT aneurysm. A filling defect was seen within the silhouette of the aneurysm, which made us suspect the presence of a clot within the aneurysm (Fig. 1b and c). The RVOT obstruction was present both at the level of the pulmonary annulus and the sub-valvar area due to compression by the thrombus within the RVOT aneurysm. The main and branch pulmonary arteries were of adequate size. The RV pressure was 130/4 mmHg and pulmonary artery pressure was 20/8 mmHg. The patient was planned for a surgical excision of the RVOT aneurysm and for a pulmonary valve replacement.

The patient underwent redo sternotomy and release of adhesions. The patient was found to have a 10 × 8 cm RVOT aneurysm. Aortic and two-stage cavo-atrial cannulation was performed. On cardiopulmonary bypass, the RVOT aneurysm and the pulmonary arteries were dissected.

Heart was arrested using antegrade, cold blood, root cardioplegia. The RVOT aneurysm was incised, and a 6 × 4 cm organized clot was found in the aneurysm sac (Fig. 2). The thrombus was evacuated and the RVOT pericardial patch was completely excised. The pulmonary annulus was found to be intact and small, and the main pulmonary artery was of adequate size. The pulmonary valve was tricuspid with dysplastic leaflets. There were no obstructive bands seen in the RVOT. The pulmonary valve leaflets were excised, and a 21 mm porcine Medtronic Hancock bioprosthesis (Medtronic Inc., Minneapolis, MN, USA) was placed in the anatomical position in the standard fashion. The anterior two-thirds of the pulmonary valve annulus and the RVOT were reconstructed using a Gore-Tex (W.L. Gore & Associates, Flagstaff, AZ, USA) patch. The patient was weaned off cardiopulmonary bypass in sinus rhythm with 5 µg/kg/min of dopamine. The postoperative course was uneventful. Postoperative echocardiogram showed normal functioning pulmonary bioprosthesis with a mean gradient of 7 mmHg. The patient was discharged on postoperative day 8 and is doing well on 3-month follow-up.

COMMENT

Trans-atrial, trans-pulmonary repair of tetralogy of Fallot has gained popularity to avoid the adverse effects of right ventriculotomy. Although it may not be possible to avoid a
ventriculotomy in all patients, the length of the ventriculotomy should be limited.

Development of RVOT aneurysms is one of the complications of ventriculotomy during repair of tetralogy of Fallot. Use of large pericardial patches to reconstruct the RVOT, residual RVOT obstruction and residual shunts are some of the common causes of RVOT aneurysms reported in the literature [3]. The use of a transannular patch, an RVOT patch or the avoidance of a patch during the reconstruction of the RVOT does not significantly change the incidence of RVOT aneurysms in the postoperative period [4].

In our patient, the presence of residual RVOT obstruction at the pulmonary annulus led to the aneurysm dilatation of the RVOT patch. Autologous pericardium has been extensively used in surgery due to its durability, tensile strength and low thrombogenicity, and we found no case reports of thrombus formation with its use. Pericardium treated with glutaraldehyde is considered less prone to dilatation [5]. The stasis within the redundant aneurysmal sac due to distal obstruction probably led to thrombus formation. We found no case reports of thrombus within an RVOT aneurysm reported in the literature.

The presence of residual RVOT echocardiographic gradient of >50 mmHg and RVOT obstruction with progressive dilatation of the RV with RV dysfunction are indications for re-operation in patients after tetralogy of Fallot repair [6]. As the pulmonary valve annulus was small and the patient had developed RV dilatation and dysfunction, we decided to do a pulmonary valve replacement. After pulmonary valve replacement, freedom from re-operation at 10 years is about 70–86% [7]. The results with both pericardial and bovine bioprosthetic valves are similar [8].

RVOT aneurysms have received little attention in recent literatures. Progressive dilatation of the RVOT aneurysm has been described as an indication for re-operation and resection of the aneurysm [3]. The presence of a thrombus within the aneurysm has the potential to embolize the pulmonary circulation. Patients with an RVOT aneurysm should be specifically evaluated for the presence of a clot within the aneurysm sac. We believe that in all RVOT aneurysms containing a clot, early surgery should be considered to prevent fatal complications.

Conflict of interest: none declared.

REFERENCES

