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Case Report

Tetralogy of Fallot with Scimitar syndrome in VACTERL: a very rare association

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ABSTRACT

The most common congenital cardiac malformation that occurs in VACTERL association is ventricular septal defect. Scimitar syndrome is a very rare congenital anomaly of the heart occurring due to maldevelopment of the lung bud or the pulmonary vascularization during early embryogenesis. The occurrence of Scimitar syndrome in VACTERL and its association with another cardiac congenital anomaly such as Tetralogy of Fallot (TOF) is extremely rare. The association of Tetralogy of Fallot with Scimitar syndrome alters the management strategy. Hereby, a very rare combination of Scimitar syndrome and tetralogy of Fallot in a four-year-old boy with VACTERL is presented, who underwent a successful single-stage surgical correction.

Keywords: Anomalous drainage, Dextroposition, Embolization, Scimitar syndrome, Tetralogy of Fallot, VACTERL

INTRODUCTION

VACTERL association is a non-random association of birth defects characterized by anomalies of the vertebra, anal atresia, cardiac malformations, tracheoesophageal fistula, renal anomalies and limb defects. A secure definition of VACTERL association includes presence of at least one anomaly in all three involved body parts (i.e. limbs, thorax and pelvis/lower abdomen), and probable definition is presence of two or more anomalies in two body parts. The most common cardiac anomalies seen in VACTERL are ventricular septal defect (VSD), atrial septal defect and tetralogy of Fallot (TOF).[1-4] Although Scimitar syndrome (cardiac malformations) represents one of six component features of the VACTERL association, it is possible that this association might have been overlooked. Our patient presented with an extremely rare combination of TOF and Scimitar syndrome in VACTERL association. This combination has not yet been reported in the literature and it is proposed that this combination is not a random occurrence but a further expansion of VACTERL association.

CASE REPORT

A four-year-old boy with VACTERL association: anal atresia, single right kidney and congenital heart disease underwent colostomy for anal atresia during the neonatal period. Cardiac evaluation revealed situs solitus, levocardia, dextroposition of the heart, hypoplastic right lung with TOF and Scimitar syndrome. There was a large subaortic maligned VSD with severe infundibular and valvular pulmonary stenosis. Pulmonary valve annulus and left pulmonary artery were good sized. Right pulmonary artery was uniformly hypo plastic. There was a large ASD shunting bidirectional, predominantly right to left with dilated right heart chambers. The right sided pulmonary veins were joining to form a scimitar vein and draining anomalously in to right atrium at the junction of inferior vena cava and right atrium. There was large aorto-pulmonary collateral from the abdominal aorta arising at the level of celiac axis and supplying the entire right lung. Preoperative assessment revealed a well-functioning colostomy and normal renal function. He underwent a successful single stage complete surgical

correction with VSD closure, infundibular resection, pulmonary valvotomy, rerouting of right sided pulmonary veins to left atrium and ligation of the collateral at the level of diaphragm. His postoperative recovery was uneventful.

DISCUSSION

Scimitar syndrome is a rare variant of partial anomalous pulmonary venous connection. The occurrence of Scimitar syndrome in VACTERL and its association with another cardiac congenital anomaly such as TOF is extremely rare.[5-8]

In the present case, we observed that the conventional management of Scimitar syndrome was altered by the association of TOF with severe infundibular pulmonary stenosis. The child was severely cyanosed with frequent squatting episodes.

The abdominal collateral could not be embolized preoperatively as the infundibular stenosis was very severe and it was the only source of pulmonary blood flow. A single stage surgical correction of both malformations was done through median sternotomy and the abdominal aortic collateral was surgically ligated at the level of the diaphragm.

In children with Scimitar syndrome there are varying degrees of dextroposition of the heart. In our patient, as the heart was dextro-rotated, the right ventricular outflow tract was more anterior than usual. In the absence of right ventricular outflow tract obstruction, a postero lateral thoracotomy would provide a direct access to the collaterals and also an easier means of dividing the anomalous vein and reimplanting into the left atrium without an intra-atrial baffle.

We chose a median sternotomy approach as it would enable more direct access to the right ventricular outflow tract, though the access to the collaterals would be difficult. The collaterals from the abdominal aorta to the right lung can branch under the diaphragm and it would be difficult to ascertain if all of them have been ligated. In the median sternotomy approach, the surgeon has direct visualization of the opening of the right pulmonary vein near the lower right atrium near the Inferior vena cava opening and the return from the opening would indicate if the collaterals have been adequately controlled.

In this patient there was a torrential return from the pulmonary veins after ligating the collaterals and hence more posterior dissection was done behind the Inferior vena cava where there were two separate branches. The pulmonary venous return was controlled after ligating the two more posteriorly located collaterals.

CONCLUSION

To summarise, the association of tetralogy of Fallot with Scimitar syndrome alters the management strategy from a staged hybrid approach to a single stage surgical management. In this rare combination of tetralogy of Fallot with Scimitar syndrome, median sternotomy would still be the preferable approach for repair. It would be ideal to embolize the collaterals by transcatheter approach before surgery. If this could not be achieved, intraoperative pulmonary venous return would be a good indicator of the adequacy of surgical collateral ligation.

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