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Brief Report

Left hemitruncus with normal right-sided pressures in an adult

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Abstract Hemitruncus is a rare congenital heart disease. Anomalous origin of the left pulmonary artery is not only rare but also pathogenetically different from anomalous origin of the right pulmonary artery from the ascending aorta. In most cases in isolated hemitrucus pressures in the right ventricle and the normally originating pulmonary artery are systemic or suprasystemic. We present a rare case of anomalous origin of the left pulmonary artery from the ascending aorta diagnosed in an adult with normal pressures in the right ventricle and normally originating pulmonary artery. To the best of our knowledge, this unique haemodynamics has never been reported in the literature.

Keywords: Hemitruncus; pulmonary hypertension; truncus arteriosus

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HEMITRUNCUS IS DEFINED AS A CONDITION IN which one branch of the pulmonary artery originates from the ascending aorta and the other branch of the pulmonary artery courses normally from the main pulmonary artery, which arises from the right ventricle.[1,2] The origin of the right or left pulmonary artery from the aorta occurs in the presence of separate aortic and pulmonary valves, and should be differentiated from the truncus arteriosus (common arterial trunk) in which the pulmonary arteries originate from the ascending aorta, but in the presence of a common semilunar valve. In isolated hemitruncus, more commonly it is the right pulmonary artery that arises anomalously from the ascending aorta and pressures in the right ventricle and the normally originating pulmonary artery are usually suprasystemic.[1,3,4]

We report a case of a 19-year-old man who was found to have hemitruncus arteriosus of the left pulmonary artery without associated intracardiac structural heart defects. To the best of our knowledge, there is no case report in the literature of anomalous left pulmonary artery arising from the ascending aorta diagnosed in an adult without associated cardiac structural defect. More so, pressures in the right ventricle and the normally originating right pulmonary artery were within normal limits, which has never been reported in the literature.

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Case report

A 19-year-old man presented with a history of easy fatigability, exertional dyspnoea and recurrent haemoptysis. On examination, he had loud wide split second heart sound and short grade 2/6 ejection systolic murmur in the left upper parasternal area. The electrocardiogram was suggestive of left ventricular hypertrophy. On chest X-ray posterior-anterior view, there was mild cardiomegaly with left ventricle type apex, and the pulmonary artery area was very prominent. An echocardiogram revealed dilatation of the left atrium and left ventricle. Pulmonary artery bifurcation was not well visualised. In high parasternal view, an anomalous vessel was seen arising from the ascending aorta. There was mild tricuspid regurgitation, and the right ventricular systolic pressure estimated by tricuspid regurgitation jet velocity was 20 mmHg. Cardiac catheterisation was done.

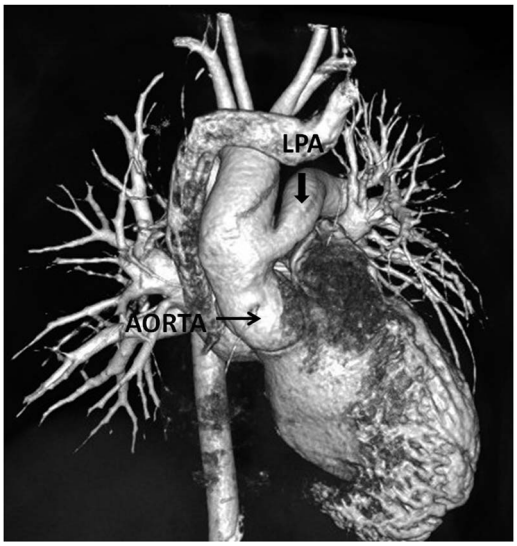

Figure 1.
Angiographic picture of the anomalous left pulmonary artery from the ascending aorta.

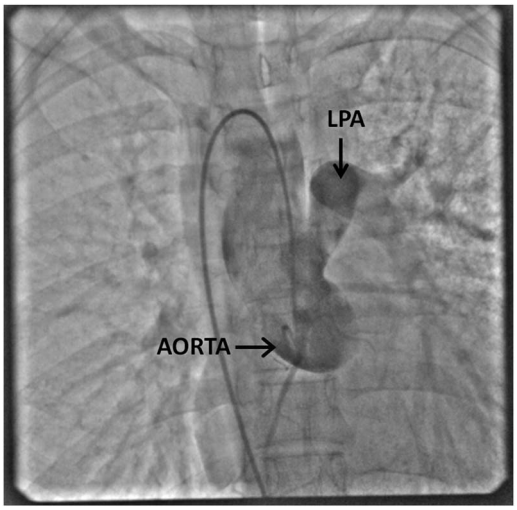

Figure 2.
Computed tomography angiographic image of the anomalous left pulmonary artery from the ascending aorta.

The right atrial mean pressure was 2 mmHg. The right ventricular systolic pressure was 18 mmHg and the right pulmonary artery pressure was 18/6 mmHg with a mean of 10 mmHg. The aortic arch was right sided. An ascending aortogram showed left pulmonary artery anomalously arising from the posterolateral aspect of the ascending aorta. The right pulmonary artery arose normally from the right ventricle. The left pulmonary artery pressure was 130/76 mmHg, same as the aortic pressure.

Discussion

Hemitruncus is best defined as a condition in which one branch of the pulmonary artery originates from the ascending aorta and the other branch of the pulmonary artery courses normally from the main pulmonary artery, which arises from the right ventricle.

It has been suggested that the anomalous origin of one pulmonary artery from the ascending aorta should be suspected when unexplained pulmonary hypertension is diagnosed and no structural intracardiac defects are observed.[3] It has been emphasised that even the lung supplied by the pulmonary artery arising from the right ventricle has high pressure and histopathological changes of pulmonary hypertension.[3–5] A crossover mechanism of a circulating vasoconstrictor or neurogenic pulmonary hypertension[6,7] from the anomalously supplied lung to the normally supplied lung was given as a possible cause of pulmonary vascular changes in the unaffected lung. More so, increased pulmonary blood flow – as the contralateral lung receives all the cardiac output in addition to the occasional blood flow of the associated anomalies, such as ductus arteriosus, aortopulmonary window, and interatrial and interventricular septal defects – has also been suggested as a critical factor in the development of pulmonary vascular disease in the normally supplied lung because patients with Tetralogy of Fallot and hemitruncus show unilateral changes only on the affected side.[4] The pulmonary vascular changes have been found at a very young age. Some studies found no significant differences between the lungs on biopsy,[7,8] whereas other studies showed more advanced changes on the non-affected side.[9,10]

Our case is unique as hemitruncus in an adult is rarely reported.[11] Another unique feature, which to the best of our knowledge, has not been reported till date is entirely normal pressures in the right ventricle and normally connected right pulmonary artery.

In a multi-centre study about the pathogenesis and associated anomalies in 108 cases, Kutche et al[1] reported that the incidence of the anomalous origin of the right pulmonary artery is not only more frequent, but is also pathogenically unrelated to the anomalous origin of the left pulmonary artery. The anomalous origin of the left pulmonary artery from the ascending aorta is an anomaly of the aortic arch, because, in all cases studied, Tetralogy of Fallot or anomalies of the aortic arch, or both, were associated. In our case also, anomalous left pulmonary artery was associated with right-sided aortic arch but without any other associated intracardiac structural heart defect.

Our case is unique because to the best of our knowledge origin of the left pulmonary artery from the ascending aorta without intracardiac structural heart defect in an adult has never been reported in the literature. Such an anomaly is usually associated with other structural defects of the heart and increased

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pressure in the normally connected pulmonary artery. In our case, the only associated anomaly was the right-sided aortic arch. The pulmonary artery pressure in the normally connected pulmonary artery was normal. The case was clinically suspected because of left ventricular hypertrophy on electrocardiogram with enlargement of the pulmonary artery on the chest X-ray, which was actually a silhouette of the anomalous left pulmonary artery, without increased right ventricular systolic pressure or shunt lesion on echocardiography.

Conclusion

In conclusion, isolated aortic origin of the left pulmonary artery without any other cardiac malformation, though a rare cardiac anomaly, can be found in an adult. It is usually associated with other arch anomalies – like right-sided aortic arch. It should be carefully searched on echocardiography when there is unexplained dilatation of the left ventricle and left atrium with prominent pulmonary artery on chest X-ray without any shunt lesion or increased right ventricular systolic pressure. The echocardiographic view for its diagnosis is high parasternal view and suprasternal view. Once diagnosed, this can be later confirmed on conventional angiography or computed tomography angiography (Figs 1 and 2).

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Conflicts of Interest

None

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