Case report

Left hemitruncus: a rare congenital heart condition

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Introduction

Hemitruncus is the anomalous origin of one of the branch pulmonary arteries from the aorta and the other arises normally from the right ventricles in the presence of 2 normal semilunar valves [1–3]. It is a rare congenital lesion with a high mortality and morbidity if not diagnosed and treated surgically early in life [1,4,5]. Usually it is an isolated anomaly, but it can be associated with other lesions as tetralogy of Fallot, aortopulmonary window, coarctation and interrupted aortic arch [1,3,5,6].

Only a few cases have been reported in the literature, most of them reporting right hemitruncus. Here we present the first case to be reported in our institute in the last 30 years. The patient was a 2-month-old baby who presented with a picture of pulmonary hypertension and was diagnosed as left hemitruncus; we discuss his presentation and diagnostic procedures.

Case report

A 2-month-old baby boy was referred to the Queen Alia Heart Institute, Amman, with a history of feeding difficulties. He had been diagnosed with double aortic arch in a private clinic and sent for surgery. On initial assessment, the physical examination showed a regular pulse of 135/min, mild chest retraction with good air entry in both lung fields. Normal first heart sound, loud second heart sound with ejection systolic murmur grade 3/6 were detected. The femoral pulses were felt.

Chest X-ray revealed increased vascularity, especially in the left lung, but there was no evidence of cardiomegaly or local lung lesion. Echocardiography revealed a dilated right heart with grade 4 tricuspid regurgitation. Doppler ultrasound investigation showed an estimated pulmonary artery pressure of > 100 mmHg, indicating severe pulmonary hypertension. Echocardiography also showed that the main pulmonary artery and the right pulmonary artery were dilated, but the left pulmonary artery was not visualized; there was no evidence of stenosis at either site. The aortic arch was seen as right arch mirror image branching pattern, with no evidence of coarctation. There was another large vessel coming off the ascending aorta having no branches, but its final course was difficult to assess with 2-D echocardiography. The right and left ventricular functions were normal and no other structural abnormalities were seen.

The patient was started on Lasix (furosemide) 1 mg/kg twice daily and Lanoxin syrup 0.005 mg/kg, within 3 days of which his symptoms had improved dramatically.

A computed tomography (CT) scan was done to better evaluate the arch; this revealed a large posterior vessel coming off just above the level of the aortic sinuses and going to the left side of the lung, the main pulmonary artery giving rise to the right pulmonary artery only. The left pulmonary artery was not seen coming from the main pulmonary artery (Figure 1); but the report was still not conclusive and the radiologist advised another catheterization to confirm the diagnosis.

Aortic root angiography was done by catheterization, and this confirmed the diagnosis of left hemitruncus (Figure 2).

The patient had no other congenital heart abnormalities. CATCH 22 syndrome was ruled out by chromosomal analysis.

The patient was sent for surgery: the left pulmonary artery was removed from the ascending aorta and reimplanted directly to the main pulmonary artery.

The patient was discharged home on Lasix (furosemide) and Lanoxin 1 week after the surgery.

At the follow-up visit 4 weeks later, the echocardiogram showed only grade 2 tricuspid regurgitation, with an estimated pulmonary artery pressure of 55 mmHg and no evidence of stenosis at the left pulmonary artery.

Discussion

Hemitruncus is a rare anomaly first described by Fraentzel in 1868 [7]. The largest series and review of this lesion was published by Kutsche and Van Mierop in 1988. This summarized a total of 108 cases, 89 with anomalous right pulmonary artery and 19 with anomalous left artery [8]. It is important to note that although left hemitruncus is less common than right hemitruncus, the former lesion is more commonly associated with either tetralogy of Fallot or right aortic arch, as in our patient who had a right aortic arch. It is intriguing that the

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Received: 16/09/12; accepted: 30/12/12

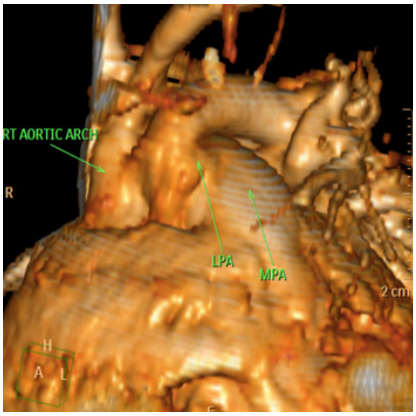

Figure 1 Computed tomography scan reconstruction, posterior view of heart, showing the aortic arch as right arch, dilated main pulmonary artery (MPA), and the large vessel coming from the ascending aorta as the left pulmonary artery (LPA)

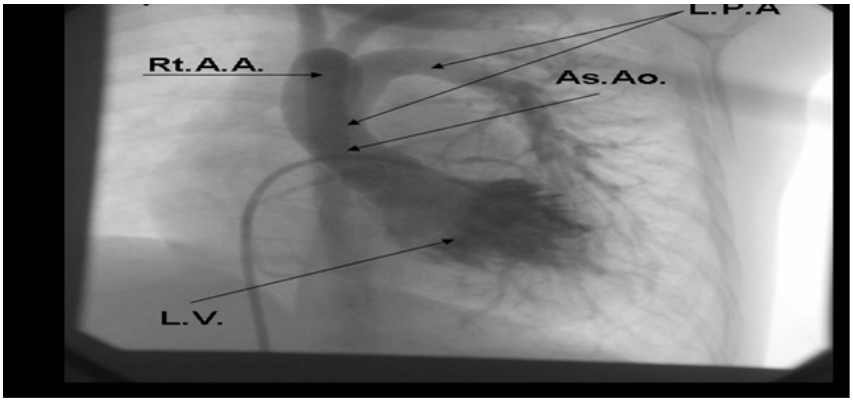

Figure 2 Angiogram of the left ventricle (L.V.) showing the right aortic arch (Rt.A.A.) with the large vessel coming off the ascending aorta (As.Ao.) as left pulmonary artery (L.P.A.)

anomalous origin of the pulmonary artery was on the opposite side of the aortic arch, as observed in this case. A patent ductus arteriosus is noted in only 13% of patients with anomalous left pulmonary artery but is seen in 69% of patients with anomalous right pulmonary artery [7–9]. In our review of the literature; although both lesions involve abnormal development of the region of the aortic and pulmonary roots and the aortic arch, they seem to have different embryologic etiologies. It is believed that the anomalous embryonic origin of the right pulmonary artery results from incomplete or delayed leftward migration of the right sixth arch [9,10]. The anomalous origin of the left pulmonary artery, in contrast, is thought to result from failure of development of the left sixth arch and persistence of the left fifth arch [11].

Early repair of this lesion is important to improve survival, which has been reported to be 30% if left untreated [6,9,12]. If left untreated, the pulmonary bed is vulnerable to early onset of pulmonary vascular obstructive disease owing to the large blood supply to both lungs for 2 reasons: a) because it receives blood at systemic pressure from the aorta and b) because it receives the entire right ventricular cardiac output unless it is protected by significant pulmonary stenosis [6,12,13].Therefore, we believe that early suspicion of this anomaly based on clinical and echocardiographic evidence was helpful in contemplating early repair in our patient and for providing appropriate counselling for the family at the time of diagnosis.

Conclusion

We conclude that this rare but serious condition is amenable to surgical repair, particularly if operated on early in life. Each patient should be evaluated by history, physical examination and the required imaging in order to reach a diagnosis that matches all the information collected.

Acknowledgements

We would like to express our gratitude to Dr Fakhri Al-Hakim of the Department of Cardiology, Royal Medical Services for all his efforts, support and contribution in reaching the final diagnosis.

Funding: None.

Competing interests: None declared.

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