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SHORT COMMUNICATION

Balanced double aortic arch with tetralogy of Fallot

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KEYWORDS

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Abstract We present a rare case of balanced double aortic arch in a 19 year old cyanotic boy, a known case of tetralogy of Fallot. This was suspected on chest X-ray and confirmed on further imaging by cardiac catheterization and computed tomography. We discuss the clinical and surgical implications of this condition.

1. Introduction

Double aortic arch is the most common form of complete vascular ring, encircling both the trachea and esophagus, resulting in non-cardiac morbidity like stridor, dyspnea, cough, recurrent respiratory tract infection and dysphagia 1. We report a case of adult asymptomatic balanced type of double aortic arch in a patient of tetralogy of Fallot who underwent successful intracardiac repair.

2. Case report

A 19 year old male patient, a known case of tetralogy of Fallot who had undergone Blalock–Taussig (BT) shunt at 4 years of age, presented with progressive complaints of exercise intolerance, fatigue and increasing cyanosis. He was referred for preoperative evaluation for corrective surgery. Physical examination revealed deep cyanosis (SpO_2 78%), grade III clubbing and an ejection systolic murmur at pulmonary area. No continuous murmur was heard. Chest X-ray suggested the presence of double aortic arch (DAA) due to indentation of bilateral aortic knobs (Fig. 1: Panel A). Echocardiography revealed large ventricular septal defect along with severe infundibular and valvular pulmonary stenosis with confluent good sized pulmonary artery and adequate sized left ventricle. BT shunt was not visualized. We could not clearly discern the type and side of arch on echocardiography. The presence of DAA was confirmed on cardiac catheterization (Fig. 1: Panel B). Computed tomography also revealed two arches of equal size, both patent, arising from ascending aorta, giving rise to corresponding subclavian and carotid arteries and merging posteriorly to form descending thoracic aorta (Fig. 2: Panel A and B). BT shunt was not visualized suggesting its occlusion. Patient subsequently underwent intracardiac repair with ligation of posterior aortic arch. Post-operative recovery was uneventful.

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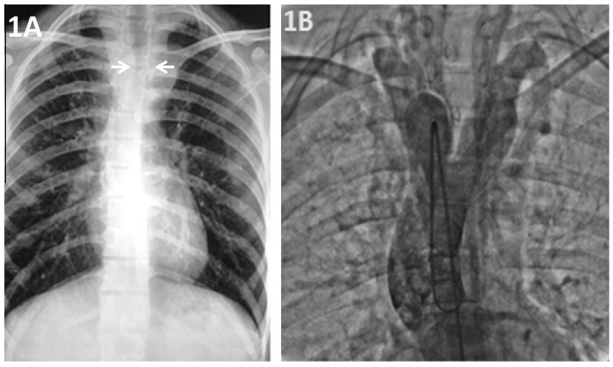

Figure 1 Panel A: Chest X-ray PA view showing double aortic arch due to bilateral aortic knobs and indentation of trachea on both sides (arrow) Panel B: Aortic root angiogram showing the presence of double aortic arch with two arches of equal sizes giving rise to ipsilateral arch vessels.

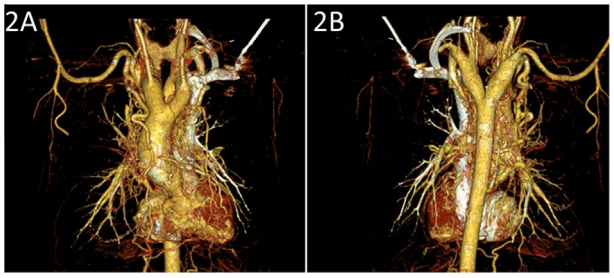

Figure 2 Panel A – (anterior view) and Panel B (posterior view):- CT angiogram showing two arches of equal size, both patent, arising from ascending aorta, giving rise to corresponding subclavian and carotid arteries and merging posteriorly to form descending thoracic aorta.

3. Discussion

These double aortic arches are classified among vascular rings that are abnormalities of the aortic arch that partially or completely encircle the esophagus and/or trachea and may cause dysphagia and/or respiratory symptoms.[1] Asymptomatic double aortic arch with tetralogy of Fallot is extremely rare.[2] The International Congenital Heart Surgery Nomenclature and Database Committee have classified double aortic arch into three groups based on dominance: right aortic arch, left aortic arch or balanced arches. Balanced arch as in the present case is least common.[3] The evaluation of the patient with suspected double aortic arch includes conventional chest X-ray in posteroanterior and lateral position; it is also possible to include esophagogram as a complementary evaluation.[4] Radiological signs suggestive of vascular ring are: mediastinal widening, presence of an aortic knob on the right, and evidence of tracheal and/or esophageal compression.[4] However, further confirmation by other imaging modalities is essential. Echocardiogram is useful to evaluate the aortic arch and heart anatomy. It is noninvasive, readily available and allows accurate delineation and exclusion of other major cardiac pathologies. However, in adult patients with poor echo window, the type and side of arch are often missed like in the present case. Supra-sternal echocardiography view most of the times provides accurate information.[5] Currently, CT and magnetic resonance (MR) imaging showed more accuracy for the evaluation of mediastinal structures.[6] They provide excellent preoperative definition with no discrepancies with surgical findings.[5] The symptoms in such patients derive from either double aortic arch (compression of the esophagus and trachea and consist of respiratory difficulties, cyanosis (specifically related with eating), stridor, and dysphagia) or from the basic intracardiac anatomy.[7] Patient in the present case did not have any symptoms due to double aortic arch but was probably symptomatic due to blocked BT shunt and hence destauration. If symptoms are present due to DAA, they often determine the course of treatment. Surgical repair in the present case was performed after taking consent from the patient and posterior arch was ligated. This was done to leave the supply of the corresponding subclavian and carotid arteries. Post operative course of patient was uneventful.

4. Conclusion

Asymptomatic balanced type of double aortic arch with tetralogy of Fallot is extremely rare. High index of clinical suspicion supplemented with stepwise radiological approach can clinch the diagnosis in most cases. This prevents the early and long-term respiratory and gastrointestinal complication. Outcomes are excellent after repair of DAA in most centers.

Conflict of interest

None declared.

Acknowledgment

None.

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