Case report

Aneurysmally dilated major aorto-pulmonary collateral in tetralogy of Fallot

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KEYWORDS

Cardiac catheterisation
Complications of cyanotic heart disease
Congenital heart disease in adults

ABSTRACT

Haemodynamically significant systemic-to-pulmonary artery collaterals may present as a cause of cardiorespiratory compromise in tetralogy of Fallot (TOF) with pulmonary atresia. We present here a case of TOF with pulmonary atresia with aneurysmally dilated aorto-pulmonary collateral causing compression over trachea, presenting as dry irritating cough who died suddenly with haemoptysis probably due to rupture of hypertrophied bronchial collaterals or direct erosion of large major aorto-pulmonary collateral arteries (MAPCA) into the respiratory tract.

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Introduction

Major aorto-pulmonary collateral arteries (MAPCAs) supply blood to the lungs when the native pulmonary circulation is underdeveloped. Instead of coming from the pulmonary trunk, supply develops from the aorta and other systemic arteries.¹ Major aorto-pulmonary collateral arteries develop early in embryonic life but regress as the normal pulmonary arteries develop.² In certain cardiac conditions, the pulmonary arteries do not develop. The collaterals continue to grow, and can become the main supply of blood to the lungs.³

Case report

We came across an interesting patient, an 18-year-old girl, symptomatic since her early childhood with cyanosis and mild effort intolerance. She presented to us with recent onset dry irritating cough and right supra-clavicular pulsatile swelling. There was no history of cyanotic spells or squatting episodes. Clinically her heart rate was 78/min, blood pressure was 124/70 mmHg, mild central cyanosis and clubbing observed. Height was 159 cm and weight 35 kg, saturation was 70% at room air and haematocrit was 40%, with body surface area of 1.28 m². There was visible pulsatile (arterial pulsations) swelling seen in right supra-clavicular region approximately

2 cm in size. Cardiovascular examination revealed single second heart sound, aortic ejection click, and continuous murmur, which was heard all over supra-sternal area, right supra-clavicular, and back.

Trans-thoracic echocardiography (TTE) was consistent with diagnosis of tetralogy of Fallot (TOF) with pulmonary atresia. Additionally, there was severely dilated branch of aorta, probably arising from arch of aorta, proximal to the right innominate artery. However, it was difficult to differentiate whether it was the native arch vessel or a MAPCA. Hence, she underwent cardiac catheterisation which revealed presence of giant MAPCA (25 mm) arising from arch of aorta, proximal to the right innominate artery supplying hilar pulmonary arteries like a natural Blalock–Taussig shunt. Pulmonary arteries were confluent and were of adequate size (9.5 mm each) (Nakata index 116/Mcgoon index 1.40) and no additional significant MAPCAs were demonstrated (Figure 1). She opted for conservative management considering the major risk involving during the corrective surgery. However, a month later she suddenly died following a bout of massive haemoptysis.

Discussion

Major aorto-pulmonary collaterals are found in about 35–40% of patients with TOF with pulmonary atresia. An inverse relation was observed between the total number of aorto-pulmonary collaterals and the size and existence of central pulmonary arteries.⁴

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Figure 1 Major aorto-pulmonary collateral arteries (MAPCAs) angiogram illustrates the huge MAPCA arising from arch of aorta and adequately sized confluent pulmonary arteries.

Our patient was presented with pressure symptoms in the form of dry irritating cough due to aneurysmally dilated MAPCA.

There is only a single case report in the literature presenting with such a size of MAPCA causing pressure symptoms. The site of origin of MAPCAs are from the descending aorta (70%), the branch of aortic arch (15–20%), and the ascending aorta (10–15%).⁵ In our case, MAPCA was observed arising from the arch, as seen in approximately 15–20% of the cases.

However, three types of systemic collateral arteries were distinguished by their origin and a characteristic type of ‘anastomosis’ with a pulmonary artery by Rabinovitch et al.⁶ Type 1 the bronchial artery branches with intrapulmonary anastomoses, type 2 direct aortic branches with hilar anastomosis as seen in our case and type 3 indirect aortic branches with extra pulmonary anastomosis.

These aorto-pulmonary collaterals can sometimes lead to pressure symptoms on account of their size as described by Comerçi et al. (aorto-pulmonary collaterals causing oesophageal indentations).⁷

Best modality of treatment in our patient would have been unifocalisation of huge MAPCA to pulmonary artery and left Blalock–Taussig shunt at the same time. Considering the proximity of important arch vessels to the aneurysm, high likelihood of thromboembolic episode and major bleeding

during the corrective surgery, conservative management was suggested.

Management was discussed with the surgeon. After explaining pros and cons of such a high-risk procedure to the relatives, they opted for medical management. Unfortunately, she died at home following about of massive haemoptysis. Autopsy could not be performed. The possible cause could be rupture of hypertrophied bronchial collaterals or direct erosion of large MAPCA into the respiratory tract. There was an identical case report describing death due to mediastinal haemorrhage likely secondary to rupture of MAPCA described by Miyazaki et al.⁸

In conclusion, we should have high index of suspicion of aneurysmally dilated MAPCA in a patient of congenital cyanotic heart disease presenting with supra-clavicular pulsations and pressure symptoms.

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