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Pulmonary atresia with ventricular septal defect type C surviving into adulthood without intervention: A case report

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Abstract

Pulmonary Atresia with Ventricular septal defect is a rare anomaly and surviving into adulthood without any surgical intervention is extremely rare. Most of the patients succumb to congestive heart failure early in life. We report a case of pulmonary atresia ventricular septal defect type C surviving at 21 years of age without prior surgical intervention.

Keywords

pulmonary atresia ventricular septal defect; pseudo truncus arteriosus; truncus arteriosus type 4.

Abbreviations

PA-VSD: Pulmonary atresia with ventricular septal defect; Tet-PA: Tetralogy with pulmonary atresia; MAPCAs: Major aortopulmonary collateral arteries; CTPA: Computed tomography pulmonary angiography.

Introduction

Pulmonary atresia with ventricular septal defect (PA-VSD) is a complex and extremely heterogeneous cardiopulmonary Malformation [1]. Initially classified as truncus arteriosus type 4, but now uniformly considered to be a form of PA-VSD [2]. Baltimore-Washington Infant Study recorded 4390 infants with cardiovascular malformations from 1981–1989. Tetralogy of Fallot were 296 (6.7%). Sixty of 296 (20%) infants in the Tetralogy group were tetralogy with pulmonary atresia (Tet-PA). Tet-PA accounted for 1.4% of all forms of congenital heart disease and 0.07 per 1000 live births [3]. Many authors have stated that patients with pseudo truncus arteriosus usually die during infancy and rarely survive to adult life [4]. Hence we would like to present a case of PA-VSD diagnosed in adulthood.

Case Presentation

A 21-year-old female presented with a history of exertional breathlessness of 2-year duration, there was no history of orthopnoea, syncope, palpitation, chest pain. There was no previous history of similar complaints and childhood was uneventful. she was recently treated for pulmonary tuberculosis.

On examination revealed grade 2 pan digital clubbing and cyanosis. The resting oxygen saturation was mildly reduced at 90% and a loud second heart sound on auscultation.

Transthoracic 2D echo revealed a single large malaligned non-restrictive Ventricular Septal Defect (VSD), overriding of aorta. Due to poor acoustic window, adequate imaging of pulmonary outflow tract could not be obtained. For better delineation of the pulmonary arterial supply, a computed tomography pulmonary angiography along with aortogram was planned.

CTPA revealed absent main pulmonary artery, right ventricular outflow tract indicating pulmonary atresia. Right Aortic Arch, large arterial channels representing MAPCAs were seen arising from the descending thoracic aorta supplying blood to lungs (Figure 1). Also seen was a large subaortic VSD (Figure 2), overriding of aorta, right ventricular hypertrophy. The absence of the native pulmonary vascular channels with MAPCAs supplying the lung directly indicates a type C PA-VSD.

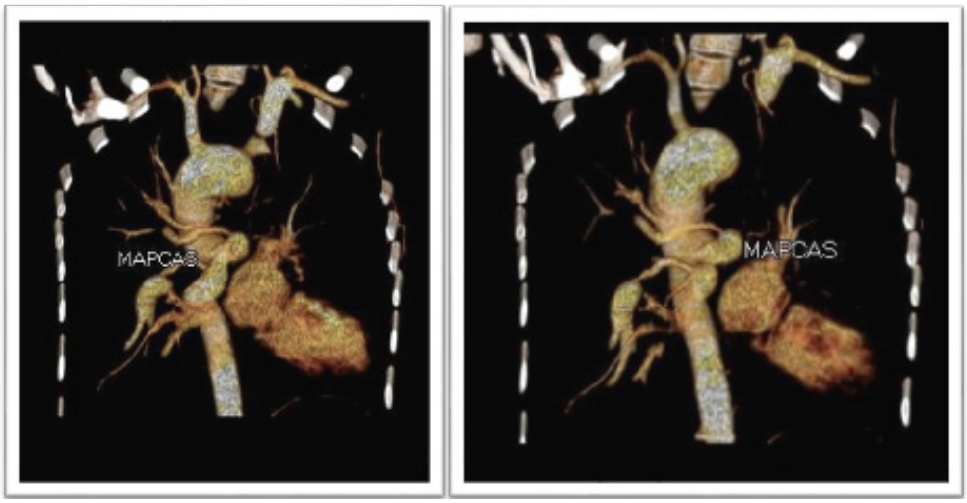

Figure 1: Coronal section of CTPA and aortogram revealing MAPCAs arising from descending thoracic aorta.

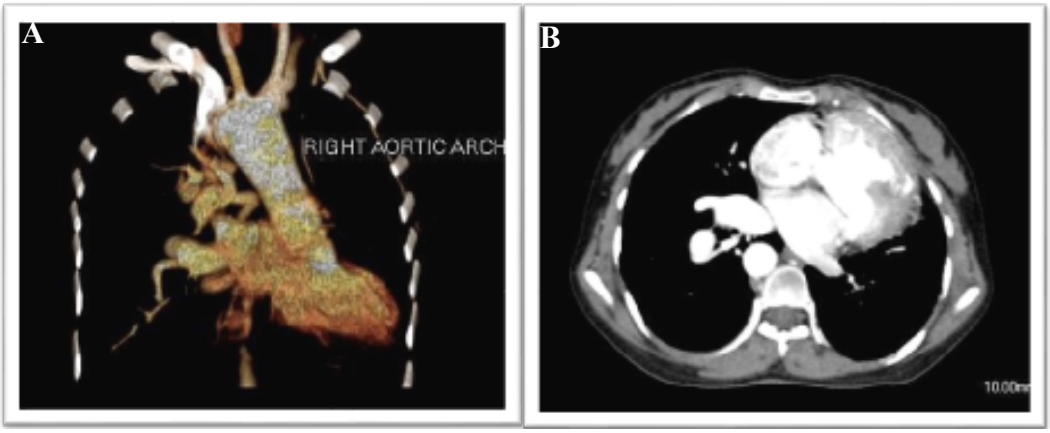

Figure 2: CCT Images A: Coronal section showing right aortic arch; B: transverse section showing large VSD.

Discussion

PA-VSD is also known as “pseudo truncus arteriosus” or “truncus arteriosus type 4” or Tet-PA [1]. Genetic, environmental, and familial factors play a causative role in etiology of PA-VSD and therefore it remains multifactorial in nature. PA-VSD is classified into 3 types depending upon the source of the pulmonary circulation [1]. In type A, there is presence of the native pulmonary arteries with a PDA supplying blood to them. In type B, there is presence of both the major aortopulmonary collateral arteries and the native pulmonary arteries. In type C, only MAPCAs are seen providing the pulmonary blood supply with absent native pulmonary arteries [1]. Many authors have stated that patients with pseudo truncus arteriosus usually die during infancy and rarely survive to adult life. In Wood’s series the ages ranged from 3 to 35 years; the ages in the series of Allanby et al, ranged from 15 weeks to 12 years; East and Barnard [4] reported 2 men who lived for 33 and 20 years, respectively; Bach’s patient died of a cerebral abscess at 30 years of age Wheeler and Abbott reported on a man living to the age of 29 years [4].

Survival past 20 years is extremely rare in PA VSD without intervention, above-described case is one such rare finding.

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