Challenging Combination; Pseudocoarctation with Subaortic and Subpulmonic Stenosis

İddialı Kombinasyon; Psödokoarktasyon ile Subaortik ve Subpulmonik Stenoz

Valvuler Stenoz ve Psödokoarktasyon / Valvuler Stenosis and Pseudocoarctation

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Özet

Kombine sol ve sağ ventrikül çıkım yolu obstrüksiyonu nadir görülen bir konjenital anomalidir. Psödokoarktasyon, aortun kıvrılma veya bükülmesi olarak bilinen, aortun nadir görülen bir anomalisidir. 4 yaşında kız hasta, hastanemize yaklaşık bir yıldır devam eden dispne ve başdönmesi şikayeti ile başvurdu. Hastada ciddi kombine sol ve sağ ventrikül çıkım yolu obstrüksiyonu ve aortada psödokoarktasyon tespit edildi. Ekokardiografik incelemede sağ ve sol ventrikül çıkım yolunda 151 mmHg ve 109 mmHg gradiyent ölçüldü. Psödokoarktasyon seviyesinde anlamlı gradiyent saptanmadı. Hastaya her iki ventrikül çıkım yolu için agresif myomektomi, sağ ventrikül çıkım yolunun perikard ile rekonstrüksiyonu, iatrojenik oluşan ventrikül septal defektin (VSD) PTFE yama ile kapatılması operasyonu uygulandı. Psödokarktasyona yönelik işlem uygulanmadı. Psödokoarktasyon ile ilgili farklı kardiyak anomaliler bildirilmiştir. Ancak,bildiğimiz kadarı ile kombine sağ ve sol ventrikül çıkım yolu darlığı ile ilişkili psödokoarktasyon daha önce bildirilmemiştir.

Anahtar Kelimeler

Psödokoarktasyon; Aort Stenozu; Kritik Pulmoner Stenoz

Abstract

Combined left and right ventricular outflow tract obstruction is an extremely rare congenital anomaly. Pseudocoarctation (PcoA), also known as kinking or buckling of the aorta, is an uncommon anomaly of the aorta. A 4-year-old girl presented to our hospital with progressive complaints of exertional dyspnea and intermittent dizziness for about one year. She was diagnosed with severe combined left and right ventricular outflow tract obstructions associated with pseudocoarctation of the aorta. Echocardiographic estimated peek pressure gradients across the right and left ventricular outflow tracts were 151 mmHg and 109 mmHg respectively. There was no significant pressure gradient across the pseudocoarctaion. Aggressive surgical myomectomy for both outflow tracts, combined with reconstruction of right ventricular outflow tract using pericardial patch and iatrogenic ventricular septal defect (VSD) closure using PTFE patch was performed. No intervention was done for pseudocoarctation of the aorta (PcoA). Association of PCoA with different types of cardiac anomalies has been reported. However to our knowledge, combined left and right ventricular outflow obstruction associated with PCoA has not been described before.

Keywords

Pseudocoarctation; Aortic Stenosis; Critical Pulmonary Stenosis

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Introduction

Although left and right ventricular outflow tract obstructions are among the most common congenital heart defects, the combination of both in the same patient appears to be very uncommon[1]. Pseudocoarctation (PcoA), also known as kinking or buckling of the aorta, is an uncommon anomaly of the aorta[2]. We describe a successful surgical treatment of congenital combined left and right ventricular obstruction associated with pseudocoarctation of the aorta in a 4 year old girl.

Case Report

An 4-year-old female was admitted to the cardiovascular department with one year history of exertional dyspnea,dizziness and intermittent confusion. Her family and past medical history was unremarkable. On physical examination, she was in good condition with a blood pressure of 90/60 mmHg, heart rate of 100-110 beats/min, respiratory rate of 21/min, and body temperature of 36 °C. Palpation of the chest revealed a thrill and on auscultation there was a grade 5/6 harsh, systolic ejection murmur associated with a mild diastolic murmur best heard along the left sternal border. Basic laboratory tests and cardiac enzyme levels were normal. The chest X-ray showed an increased cardiothoracic ratio and diminished pulmonary vascular markings. Electrocardiography showed sinus rhythm with biventricular hypertrophy and strain pattern. Echocardiography revealed severe hypertrophy of the left ventricle posterior wall(LV), right ventricle(RV) free wall, and interventricular septum(IVS). There were apparent muscular bundles at the level of the RVOT causing low-lying type stenosis. Estimated maximal pressure gradients across the right and left ventricular outflow tracts (LVOT) were 151 mmHg and 109 mmHg respectively (Figure 1). There was mild aortic regurgitation. Angiography demonstrated severely hypertrophied right and left ventricles, severe sub-valvular aortic and pulmonary stenosis (Figure 2). Pressure measurements revealed a peak-to-peak systolic gradient of 95 mmHg at LVOT and 140 mmHg at RVOT. In addition aortic angiogram showed severe buckling/ pseudocoarctation of distal arcus aorta. However, there was no significant pressure gradient across this area.

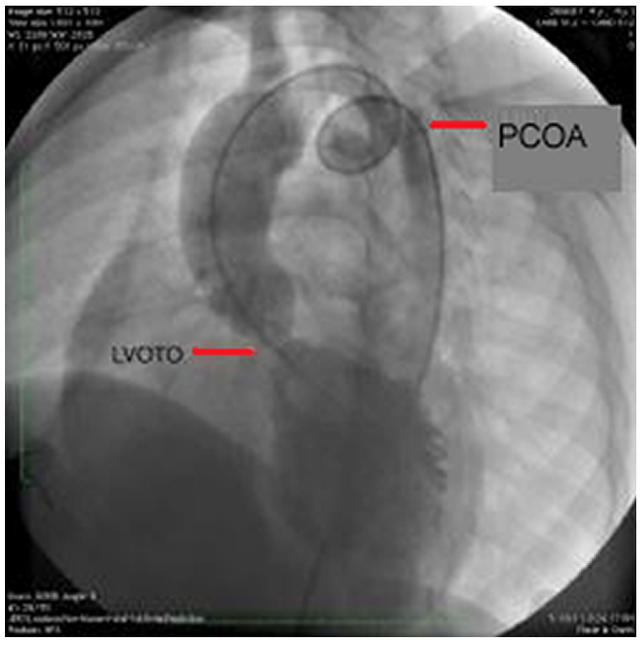

Figure 1. Angiography demonstrated complex (discrete and short tunnel type) LVOT obstruction and pseudocoarctation (PCoA)

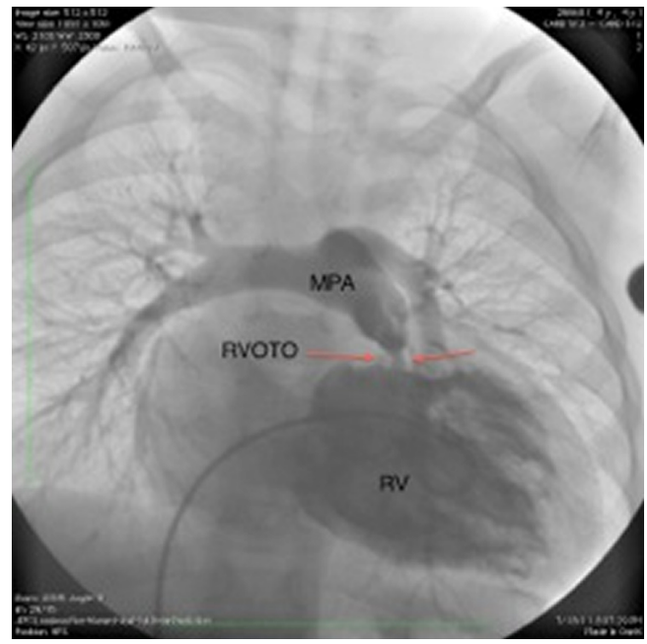

Figure 2. Angiography demonstrated low-lying type RVOT obstruction. PA annulus and pulmonary valve appeared normal.

Standard cardiopulmonary bypass and myocardial preservation techniques were used under moderate hypothermia. After aortotomy, excision of the septum was initiated with two parallel longitudinal incisions: one beneath the nadir of the right coronary cusp and the other beneath the commissure separating the left and right cusps. The mass of myocardium extending to the posteromedial papillary muscle was then excised. There wasn't enough LVOT opening by observation so additional muscle mass was excised toward the lateral attachment of the anterior mitral leaflet and adequate LVOT opening was achieved. In order to perform RVOT reconstruction a 3 cm longutidinal ventriculotomy was done. After proper muscle excision both from conal septum and parietal wall. İatrogenic VSD closed using PTFE patch. After closing the aortotomy cross clemp was removed and RVOT reconstruction was completed using glutheraldehyde treated pericardial patch on beating heart. Adequacy of myectomy was confirmed through measurement of simultaneous LVOT and RVOT pressures before and after cardiopulmonary bypass. Direct measurements after cardiopulmonary bypass revealed 14 mmHg and 9 mmHg pressure gradients across the left and right ventricular outflow tracts respectively. Postoperative complete A-V block was detected with a heart rate of 80-90 beat/min. Complete atrio-ventricular heart block persisted after two weeks and an epicardial dual chambered permanent pacemaker (Medtronic®) was implanted. The patient was asymptomatic at three-months follow-up.

Discussion

Congenital valvular, sub-valvular aortic stenosis and infundibular or subvalvular pulmonary stenosis are relatively common congenital cardiac anomalies when they occur as isolated defects or in association with other intracardiac deformities. The combination of left and right ventricular outflow obstruction is extremely rare. Milazzo AS reported an infant with sub-valvular

and valvular pulmonary stenosis, sub-valvular, and valvular aortic stenosis and hypertrophic cardiomyopathy, who presented with pulmonary hemorrhage[1]. Pseudocoarctation (PcoA), also known as kinking or buckling of the aorta, is an uncommon anomaly of the aorta[2].In our patient LVOT obstruction was mainly sub-valvular, aortic annulus size was acceptable and the aortic valvular insufficiency was mild. For this reason we performed a very aggressive circumferential myomectomy to relieve the obstruction. Occurrence of third-degree complete A-V block and iatrogenic VSD undoubtedly is the result of very aggressive surgical approach in order to remove the LVOT obstruction. An aggressive circumferential myomectomy was performed . Valeske et al. reported that AV block was most likely related to the type of LVOT obstruction. In short discrete lesions the incidence of permanent complete AV block was none. However in complex LVOT obstruction like in our patient the incidence was up to 23%[2]. A variety of abnormalities were reported in association with pseudocoarctation, including bicuspid aortic valve, aortic stenosis, ventricular septal defect, atrial septal defect, patent ductus arteriosus[3]. PCoA is different from true coarctation in the following aspects (a) Unlike true coarctation, in which the aortic arch does not reach clavicle, the aortic arch of pseudocoarctation is elongated (b) Absence or mild degree of stenosis of aortic lumen. (c) No or small blood pressure gradient across the lesion ($\Delta p < 25$ mmHg). (d) Absence of collateral circulation[4]. PCoA used to be considered a benign lesion requiring no treatment. Steınberg et al reported that congenital aortic stenosis and pseudocoarctation with four cases in autopsy series[5]. In our patient hemodynamic data and angiography along with physical examination correlated well with these rules. Since the pressure gradient across the PcoA was 10 mmHg and the patient was normotensive, no intervention was needed. Many reports showed that combination of PcoA with cardiac anomalies but to our knowledge, combined LVOT and RVOT obstruction with PcoA has not been described before.

Competing interests

The authors declare that they have no competing interests.

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