

Extrinsic Compression of Left Main Coronary Artery from Aneurysmal Dilation of Pulmonary Trunk in an Adolescent. Involution after Surgery Occlusion of Sinus Venosus Atrial Septal Defect and Pulmonary Trunk Plasty for Reduction

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We report the case of an adolescent referred with initial diagnosis of pulmonary hypertension. Non-invasive investigation disclosed a sinus venous atrial septal defect with pulmonary hypertension. The hemodynamic study confirmed diagnosis, and also showed extrinsic compression of left main coronary artery by pulmonary trunk. Surgical closure of the defect in addition to pulmonary trunk plasty were undertaken. Two years after the surgery the patient is well, with clinical signs of mild pulmonary hypertension, and showing no evidence – also on echocardiogram – of left coronary artery trunk obstruction.

Introduction

Extrinsic compression of left coronary artery (LCA) by pulmonary artery trunk (PAT) is rare, and usually associated to IAC and/or pulmonary hypertension¹⁻⁵. Even more rarely it is associated to persistence of arterial canal⁶, tetralogy of Fallot (TOF)⁷, and atrioventricular septal defect⁸. Most studies that have been published report on adult patients that may present precordial pain and myocardial ischemia, requiring revascularization simultaneously to the correction of defect that causes PAT dilation^{1,4,6}. We report the case of an adolescent with sinus venosus atrial septal defect (ASD), with proximal compression of LCA, with uneventful evolution after the occlusion of defect and PAT reduction plasty.

Case Report

A 16-year-old adolescent reporting complaints of stress dyspnea and atypical, occasional precordial pain was referred to the Clinics Hospital, at Ribeirão Preto Medical School, with an initial diagnosis of primary pulmonary hypertension, and with no structural defect shown on

Key words

Pulmonary hypertension, coronary artery anomalies, atrial septal defect / surgery.

echocardiogram. The patient was in good condition at physical exam, pulse was normal, and ++/6+ systolic murmur at pulmonary focus, with no thrill. Heart sound 2 was steadily unfolded, with a hyperphonic pulmonary component. ECG showed signs of significant right ventricular overburden. Thoracic X-Ray showed pulmonary hyperflow, right branch of pulmonary artery dilation, and quite dilated PAT (Fig. 1A). Thoracic CAT scan (Fig. 1B), as well as magnetic angioresonance (Fig. 1C) pointed out the significant dilation of pulmonary branches and trunk, and relative aortic caliber decrease. Transesophageal echocardiogram showed a 10 mm sinus venosus IAC, with anomalous venous drainage of right upper pulmonary vein in right atrium. The hemodynamic study confirmed non-invasive findings and showed significant compression of LCA trunk by PAT (Fig. 1D). Aortic pressure was 126/93 mmHg, while pulmonary artery pressure was 110/70 mmHg, decreasing to 73/52 mmHg with O₂ inhalation. Systemic vascular resistance (SVR) was 13 UWood, pulmonary vascular resistance (PVR) was 6.8 UWood. Pulmonary/systemic flow ratio was 2.9. The patient was submitted to surgery on January 28, 2004. After pericardium opening, PAT diameter called the attention since it was three times larger than aortic diameter. The ASD was occluded with bovine pericardial strip, the anomalous pulmonary vein was directed into the left atrium, with reduction plasty of PAT. Immediate evolution was favorable and the patient was discharged on captopril one week after hospital admission. Six months later the patient was in good condition, and the hemodynamic study showed slight residual ASD, with pulmonary/systemic flow ratio at 1.05, PVR: 7 UWood, SVR: 13.7 UWood, pulmonary artery pressure: 75/52 mmHg, and aortic pressure: 110/83 mmHg. Extrinsic compression of LCA was also observed to have been reduced (Fig. 2A). Twenty-one months after the surgery the patient was asymptomatic, on 50 mg/day of captopril. Cardiac auscultation revealed a pulmonary click with S2 mildly hyperphonic. Significant regression of right ventricular overburden was shown on the ECG; discreet reduction of PAT was shown on the X-Ray (Fig. 2B). Transthoracic echocardiogram showed pulmonary artery systolic pressure at 55 mmHg, intact interatrial septum, right ventricle hypertrophy, no compression of LCA (Fig. 2C), and PAT still dilated (Fig. 2D). Stress echocardiogram with dobutamine was negative for myocardial ischemia.

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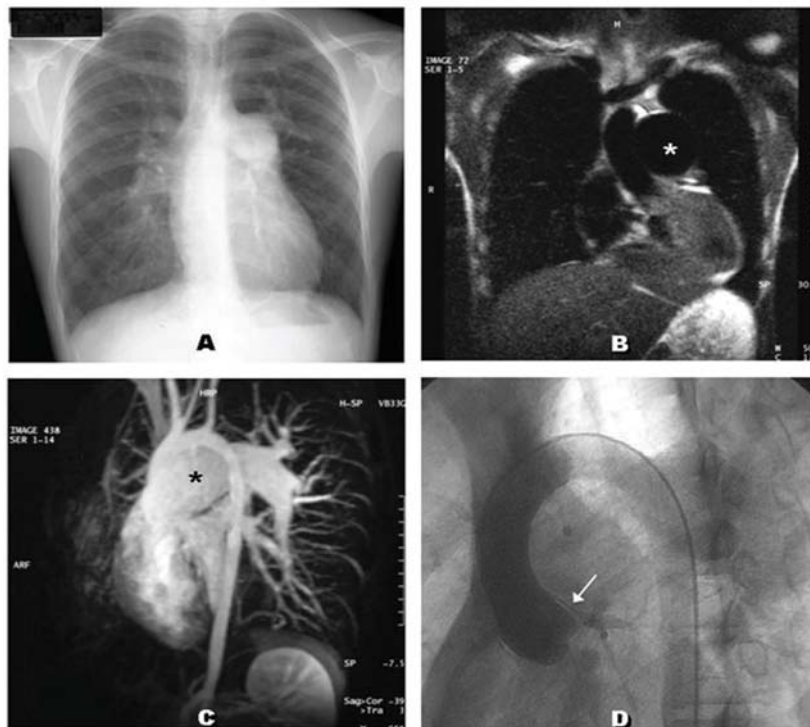


Fig. 1 - (pre-operative) - Aneurysmal dilation of PAT demonstrated by thorax X-Ray (A), CAT scan (B), and magnetic angiogram (C); (D) Hemodynamic study: aortography on left anterior oblique incidence, with evidence of significant obstruction at coronary trunk (arrow).

Discussion

Described for the first time almost half a century ago⁹, extrinsic compression of LCA by PAT is not common. Comprehensive studies are rare, although information has been available in literature on the occurrence of 2 patients (1.8%) out of 48 IAC cases studied by Kothari et al¹⁰. Another relevant study was conducted in 1989 by Mitsudo¹, and has stood as a landmark for topic discussion. While studying 38 patients in the 15-62-year-old range, the author found 7 cases (18%) of extrinsic compression of LCA. The figures reached 44% for cases with pulmonary hypertension associated.

Most cases described reveal a frequent association with IAC^{1,2,4,5,11}; however, other conditions – such as persistence of arterial canal⁶, tetralogy of Fallot (TOF)⁷, and atrioventricular septal defect⁸ – may be involved. Primary pulmonary hypertension patients with significant PAT dilation may also report the same conditions^{3,12}. Precordial pain may act as suspicion of such serious complication in patients with this type of cardiopathy and with significant PAT dilation on thoracic X-Ray. The ECG may reveal ischemic changes^{2,3,6}, and coronary obstruction may be visualized on echocardiogram and on nuclear magnetic resonance. However, diagnostic confirmation and the assessment of compression severity is usually more reliable through coronariography.

The case presented includes some characteristics which we understand justify this report. Although the topic has

been discussed based on fairly extensive studies in this country⁴, our patient seems to be the first case to be published in our journals. Additionally, the occurrence of such complication in an adolescent – as described – seems to be quite rare. To our knowledge, there is only one report in that age range¹¹.

Another interesting aspect – also made evident in our case – is the conduct adopted. Some authors believe in the need of revascularization for cases with significant obstruction¹. Others suggest that surgical reduction of PAT only may eliminate compression of the LCA, particularly if pulmonary hypertension is considered reversible². Our patient had significant signs of severe pulmonary hypertension, with good response to O₂. Almost 2 years after surgery, considerable involution of the obstruction could be seen on the echocardiogram.

To conclude, we could say that extrinsic compression of LCA by PAT is quite rare. As suggested, it may, therefore, be underdiagnosed⁴. Due to the possibility of obstruction involution, as demonstrated, many IAC cases that have been operated on without visualization of the coronaries may not have been properly documented. Cardiopathy patients that develop PAT increase (with or without pulmonary hypertension), must be carefully investigated, particularly if associated to precordial pain complaint. Due to the rare degree of the condition - which hinders any randomized therapy study – the authors find it hard to define whether routine revascularization for cases with significant obstruction

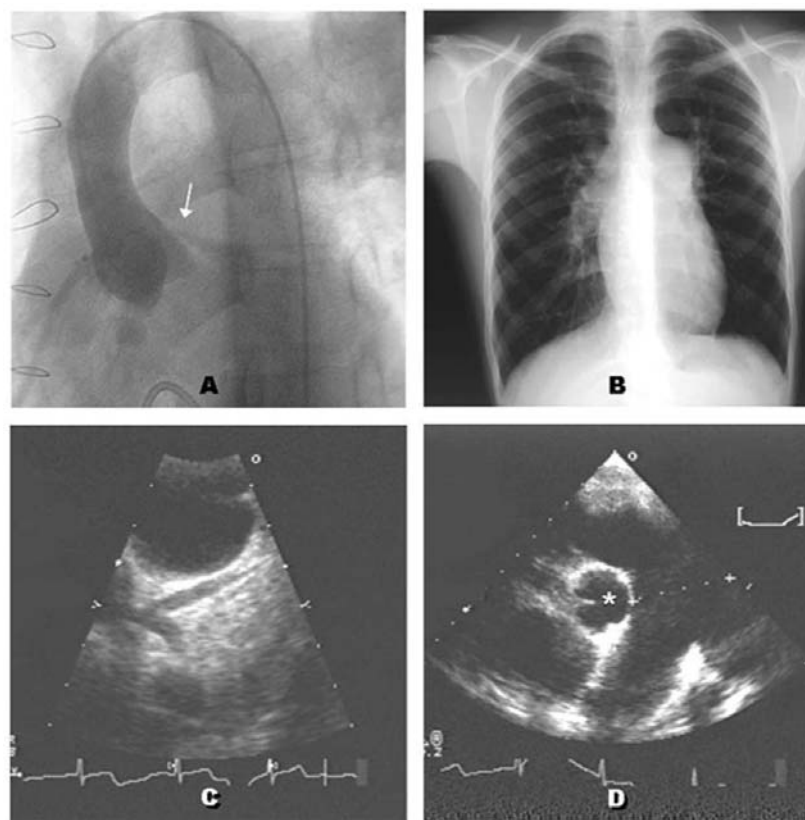


Fig. 2 - (post-operative) - A) Six months after surgery, hemodynamic study shows decrease of coronary obstruction (arrow); **B)** Two years after surgery, evaluation shows discreet decrease of PAT on thoracic X-Ray **(B)** and echocardiogram showing no evidence of left coronary artery **(C)**, although PAT still dilated **(D)**.

is required or not. However, based on the case reported here, and on the small amount of information available in the literature, it does seem that for young patients with no injury

in other arteries and with reversible pulmonary hypertension, the most appropriate conduct is correction of cardiac defect associated to PAT reduction plasty.

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