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Case Report

Transvalvular Aortico left ventricular tunnel – A rare congenital cardiac anomaly

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ABSTRACT

The aortico-left ventricular tunnel is an abnormal congenital paravalvular communication between the ascending aorta and the cavity of left ventricle. Clinical picture suggests severe aortic incompetence. We report the case of a teenage male patient whose echocardiography revealed a Type III Aortico-left ventricular Tunnel with the aortic end of the tunnel originating through a fenestration in the right coronary cusp, which is not so far reported in literature. The patient had uneventful recovery after surgical patch closure of the tunnel.

1. Introduction

Aortico–left ventricular tunnel (ALVT) is an extremely rare congenital cardiac anomaly consisting of a paravalvular communication between the aortic root and left ventricular cavity. It represents only 0.001 percent of all congenital cardiac anomalies. Approximately 130 cases have been reported in the literature, about twice as many cases in males as in females. The diagnosis of aorto-left ventricular tunnel should be considered in any infant or young child presenting with clinical findings of severe aortic incompetence.

We report a rare variant of ALVT in a teenager, who presented with clinical features similar to severe aortic regurgitation. Instead of the classical paravalvular communication, the patient had a 'Transvalvular' ALVT where the aortic opening of the tunnel was through a fenestration in the right coronary cusp of the aortic valve. Transvalvular variant of ALVT is not so far reported in the literature.

2. Case report

A 17 year old male patient presented with history of palpitation and exertional dyspnea since 5 years. There was no history of chest pain, or syncope. Clinical examination revealed an average built male who was asymptomatic at rest. Pulse was 92 bpm, high volume and collapsing in character. Hill's sign was positive with a blood pressure of 140/40 mm Hg in the right arm and 160/40 mm Hg at the thigh. Precordial

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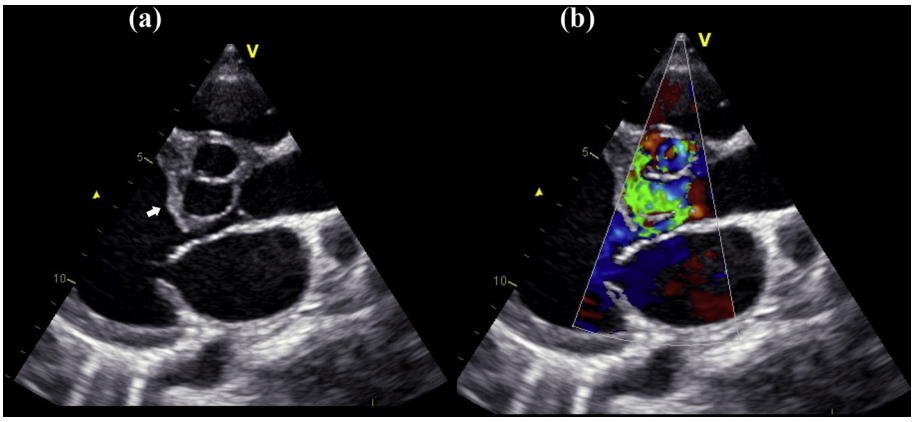

Fig. 1 – Parasternal long axis view (*a*) showing the aneurysmally dilated distal end of the tunnel in the upper part of interventricular septum. (*b*) Color Doppler showing the regurgitant jet through the tunnel into the LVOT.

examination revealed cardiomegaly, normal first heart sound, normal intensity of second heart sound and a Grade 3/6 continuous murmur along the left sternal border.

Chest X ray showed cardiomegaly with a cardiothoracic ratio of 60%. ECG revealed sinus rhythm with no evidence of chamber hypertrophy. Transthoracic echocardiogram revealed a dilated left ventricle with normal left ventricular systolic function. In the parasternal long axis (PLAX) view, an abnormal communication was noted between the aortic root, extending from the right aortic cusp, tunneling through the crest of the inter ventricular septum and draining into the left ventricular outflow tract (LVOT) (Fig. 1a &b; video 1, 2 & 3). Aneurysmal dilatation was noted at its exit site in the LVOT. In the modified PLAX view and short axis view at aortic valve level, a fenestration was noted in the right coronary cusp of aortic valve which formed the aortic end of the tunnel (Fig. 2, video 4 & 5). The lower extent of the tunnel in the ventricular septum was seen in the parasternal short axis view (Fig. 3a, video 6). Severe aortic run-off into the LVOT through the channel was noted on Doppler interrogation (Fig. 4, video 7 & 8). No windsock, dilatation or out pouching of the Sinus of Valsalva was noted. There was no evidence of any paravalvular communication also. Same findings were confirmed by a Transesophageal echo. Possibility of Sinus of Valsalva Aneurysm rupturing into the left ventricle was considered, but owing to the lack of obvious sinus dilatation and the absence of draining 'windsock', this was considered less likely. The possibility of coronary cameral fistula was ruled out as the coronary arteries appeared normal. Absence of paravalvular communication between ascending aorta and left ventricle was against the diagnosis of classical Aortico LV tunnel. Since aortic communication of the channel was through a fenestration in the aortic cusp, 'Transvalvular Aortico LV tunnel', a variant of Aortico LV tunnel was considered and the patient was referred to the Cardiothoracic Surgeon for surgical repair.

Supplementary data related to this article can be found online at http://dx.doi.org/10.1016/j.jicc.2014.12.003.

Intraoperatively, the echocardiographic findings were confirmed. A communication between the aortic root and LVOT was noted through a fenestration in the right aortic cusp which tunneled through the crest of the interventricular septum to drain in to the LVOT. The morphology suggested

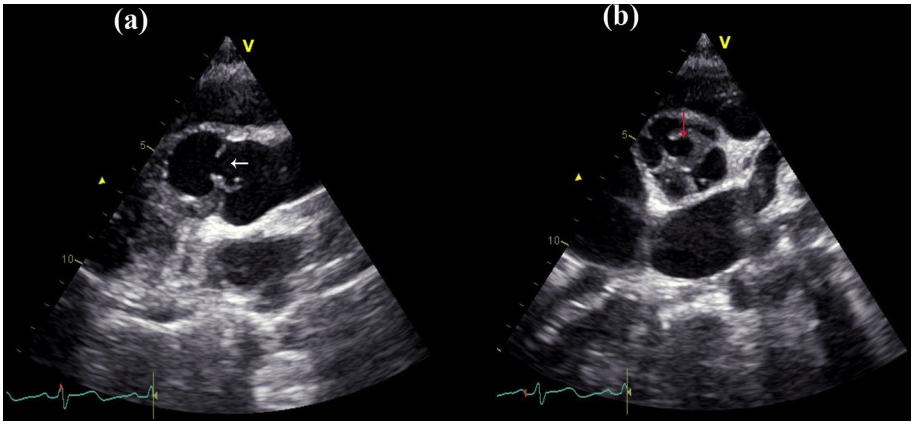

Fig. 2 – (*a*) Modified Parasternal long axis view showing the opening into the tunnel through a fenestration (arrow) in the right aortic cusp. (*b*) Parasternal short axis view showing fenestration in the right aortic cusp (arrow).