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Unusual Case of PDA with VSD in a Young Patient Developing Eisenmenger Syndrome

KEYWORDS: PDA, VSD, Eisenmenger syndrome, echocardiography, MDCT

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ABSTRACT *Both patent ductus arteriosus (PDA) and ventricular septal defect (VSD) are common congenital cardiac anomalies. But there association is relatively uncommon. Because of the existence of two separate systemic-pulmonary arterial shunts, pulmonary hypertension would be expected as a common accompaniment with developing Eisenmenger syndrome.*

CASE REPORT

A 16 yr old boy presented with anterior chest wall bulge and shortness of breath. On clinical examination, there was differential clubbing of lower limb fingers and clinically suspected pulmonary arterial hypertension (Fig 1).

Chest radiograph showed filling in of the aorto-pulmonary window and enlargement of the pulmonary arteries and cardiomegaly, especially right-sided enlargement. There is round pneumonia of the right upper lobe (Fig 2).

On echocardiography VSD and PDA were identified with tricuspid regurgitation. A dilated main pulmonary artery was also seen (Fig 3).

Multidetector-row CT (MDCT) showed presence of PDA (10 mm diameter), presence of pulmonary hypertension (aneurysmal dilatation of main pulmonary trunk >3 cm) and presence of enlarged RV and RA (Figs 4).

DISCUSSION

PDA is seen in 5 - 10% of all congenital cardiac lesions. The defect must persist beyond 3 months of age to be considered pathological.[1] Most children with PDA are asymptomatic; however, 50% of patients with a large PDA will develop pulmonary hypertension[2] and may eventually develop Eisenmenger's syndrome if left patent.

Ventricular septal defect (VSD) and patent ductus arteriosus (PDA) are common congenital cardiac anomalies occurring singly, the two combined are relatively uncommon. Because of the existence of two separate systemic-pulmonary arterial shunts, pulmonary hypertension is expected as a common accompaniment and congestive cardiac failure a common presentation in early childhood.[3] Here in our case both the conditions are present, and 16 years boy presented with features of pulmonary hypertension.

Eisenmenger syndrome is a cyanotic heart defect characterized by a long-standing intracardiac shunt that eventually reverses to a right-to-left shunt. Eisenmenger syndrome refers to the combination of a cardiac shunt (systemic-to-pulmonary), significant enough to cause cyanosis and overtime pulmonary hypertension. Patients with Eisenmenger syndrome are cyanotic and may have differential cyanosis (cyanosis and clubbing of the toes but not the fingers because the right-to-left ductal shunting is distal to the subclavian arteries). Cyanosis may be more profound when systemic vascular resistance is decreased, such as in hot weather or after exercise. There may be no murmur during systole or diastole, because shunting may be minimal.

On chest radiograph PDA complicated by Eisenmenger syndrome show filling in of the aorto-pulmonary window and enlargement of the pulmonary arteries, pruning of the pulmonary arteries peripherally, cardiomegaly, especially right-sided enlargement. The most reliable (but uncommon) sign is a calcified PDA 'railroad track' sign.[4, 5]

In our patient, Fig. 2 demonstrates an enlarged cardiac shadow with an enlarged right atrium and right ventricle. The aortopulmonary window is filled in, and there is peripheral pruning suggestive of pulmonary hypertension. There is also patchy consolidation of the right upper lobe.

Once Eisenmenger's syndrome is established, the echo will reveal pulmonary hypertension but may miss the PDA (due to the reversed flow across the shunt).[1] In our case aneurysmal dilatation of main pulmonary artery was diagnosed. He was found to have an enlarged right ventricle and tricuspid regurgitation. VSD was clearly demonstrated.

Multidetector computed tomography (MDCT) has become an important investigation in patients with PDA, especially those with associated pulmonary hypertension and Eisenmenger syndrome. Typical findings on MDCT of an uncomplicated PDA would include a tubular structure connecting the descending aorta with the distal main pulmonary artery or the proximal left pulmonary artery. Accurate measurements including narrowest and widest diameter of the PDA as well as length of the PDA can accurately and easily be done on MDCT to assist surgical management.[6] MDCT also demonstrates the extent of calcification associated with the PDA, which will influence surgical management.[7]

MDCT assists in evaluating the direction of the shunt associated with a PDA. Studies timed for the pulmonary artery will show a 'negative jet' of unenhanced blood from the aorta to the pulmonary artery via the PDA. Studies timed for the aorta will show a 'positive jet' of enhanced blood flowing from the aorta to the unenhanced pulmonary artery via the PDA.[6]

This case had all features of PDA with Eisenmenger's syndrome and also VSD. As for the present case, it was decided that PDA should be closed initially because the already existing left-to-right shunt particularly depended on PDA rather than VSD so that the closure of PDA would improve the hemodynamical features of the patient and later VSD can be repaired.[8]

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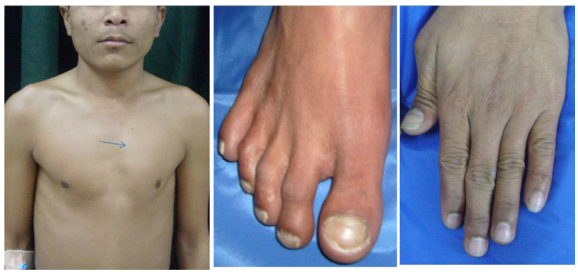

Fig. 1: clinical photograph showing anterior chest wall bulge and differential clubbing of lower limb digits.

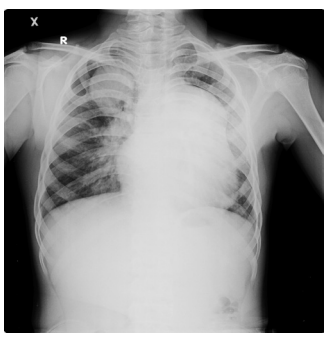

Fig. 2: chest radiograph showing dilated pulmonary artery with mild cardiomegaly. Right upper lobe round pneumonia is seen.

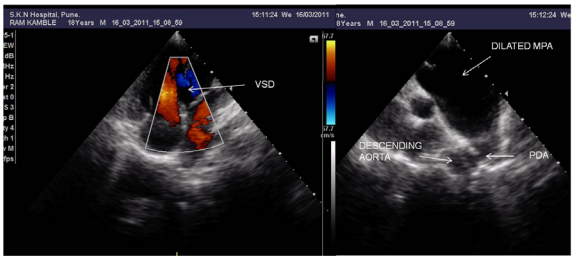

Fig. 3: echocardiogram showing VSD, dilated main pulmonary artery and PDA.

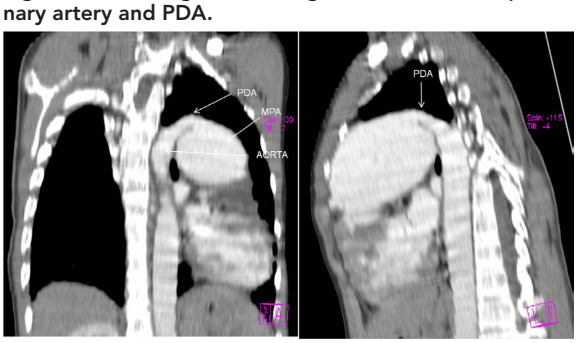

Fig. 4: MDCT reconstructed images clearly depicting PDA and dilated main pulmonary artery.

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