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Pulmonary Veno Occlusive Disease

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Pulmonary venous hypertension (PVH) is usually encountered clinically as a consequence of left ventricular failure, mitral valve disease, fibrosing mediastinitis and rarely because of pulmonary veno-occlusive disease (PVOD). The hall-marks of PVH are pulmonary congestion and edema. Pulmonary venous hypertension elicits pulmonary arterial hypertension, which may obscure PVH clinically. PVH is said to exist when pulmonary venous pressure exceeds 12 mm Hg(l). Recently, the first case of PVOD was reported from India(2). We also report our experience with a similar patient.

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Case Report

An 8-year-old boy, resident of Jodhpur, was admitted with the complaints of cough, swelling of both feet, pain in both knees and breathlessness at rest for 7 days. He was also complaining of cough, breathlessness off and on for the past 1 year with loss of weight for which he was taking treatment from a nearby dispensary. He was in good health before this.

On admission, he was pale, had edema

feet, a respiratory rate of 88/min, heart rate of 188/min, blood pressure of 100/70 mm Hg raised jugulo-venous pressure with a positive hepato-jugular reflux, hepatomegaly (liver span 14 cm) and bilateral basal crepitations. Examination of cardiovascular system revealed tachycardia, apex beat in the 5th intercostal space, medial to mid clavicular line and very loud pulmonary component of second heart sound with ejection systolic and early diastolic murmur in the pulmonary area. On the 2nd day of admission he had frequent episodes of tachy-(140/min) and bradycardia (60/min). His hemoglobin was 8g/dl. The X-ray chest revealed a cardio-thoracic ratio of 60%, prominent pulmonary arteries, right artial enlargement, interstitial haziness, prominent upper lobe pulmonary veins and Kerley B lines *(Fig. 1)*. ECG revealed an axis of +90 degree and P pulmonale in Lead II with right ventricular hypertrophy. Echocardiography showed enlarged right atrium and ventricle *(Fig, 2)* dilated pulmonary artery with normal size left atrium, ventricle, aorta and normal mitral, tricuspid, pulmonary and aortic valves. Apical 4 chamber view revealed normal opening of all the four pulmonary veins into the left atrium. There was no evidence of atrial or ventricular septal defects. Doppler echocardiography revealed peak flows at pulmonary and tricuspid valves of 69 and 60 cm/sec, respectively. The predicted pulmonary artery systolic pressure was 47 mm Hg. There was evidence of mild pulmonary and trivial tricuspid regurgitations with no Doppler signals to suggest either atrial septal defect, ventricular septal defect or patent ductus arteriosus.

A clinical diagnosis of pulmonary arterial hypertension with pulmonary regurgitation, pulmonary edema and right sided heart failur was made. The possibility of PVOD was entertained because of orthopnea, evidence of pulmonary edema and venous hypertension with normal left side heart on echo-cardiography.

The child was treated with diuretics, digoxin, corticosteroids and oral potassium supplementation. Over a period of ten days, he showed clinical as well as radiological improvement. The patient again deteriorated on day 20th and developed an increase in heart (150/min) and respiratory (70/min) rate, distress at rest, hepatomegaly and positive hepato-jugular reflux. Repeat X-ray chest PA view revealed Cardio-thoracic ratio > 60%, right atrial enlargement, interstitial haziness, prominent upper lobe pulmonary veins and Kerley B lines. The patient was again given parenteral steroids, diuretics, intravenous fluid and oxygen; but ultimately the patient absconded.

Discussion

Primary pulmonary hypertension may be idiopathic, thromboembolic or due to PVOD. These subsets are often clinically indistinguishable. PVOD is the least common of all types of unexplained pulmonary hypertension; only 30 patients of PVOD were reported in the literature till 1976(3). Predominantly children and young adults are affected, but the age has ranged from infancy to 48 years (l). Nearly one third of the cases have occurred in children. The youngest reported patient was an 8 weeks old young infant (3). The postulated etiologies are viral infection, toxoplasmosis (4), coagulation abnormalities, autoimmune mechanism (5,6), radiation(7), antimetabolites (chemotherapy), toxin and mediastinal fibrosis(l).

The lungs are the seat of congestion, edema and focal fibrosis, which may become extensive. The most striking morphological change is the inflammatory thrombotic process seen in pulmonary veins and

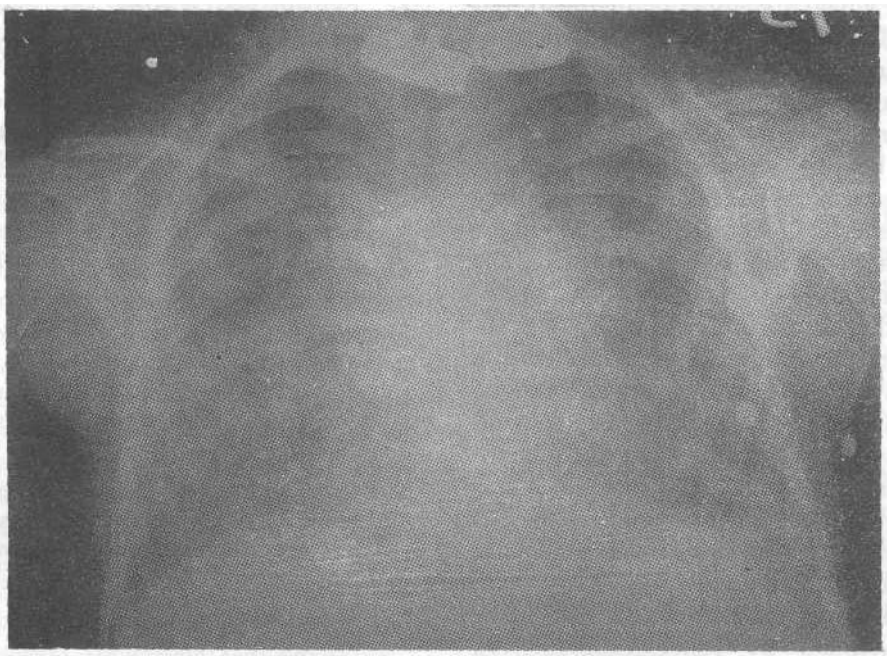

Fig. 1. X-ray chest showing cardiomegaly and prominent pulmonary vasculature with diffuse interstitial hazyiness, suggestive of pulmonary edema.

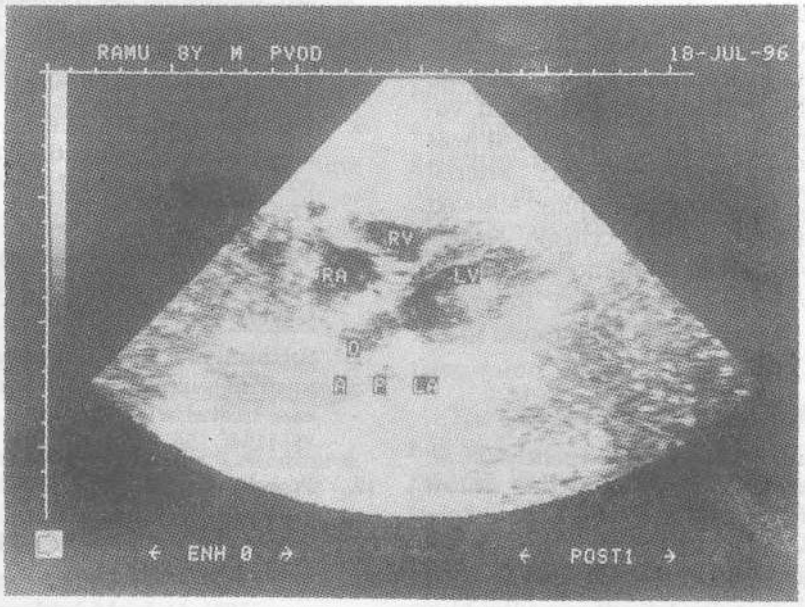

Fig. 2. Echocardiography showing enlargement of right atrium and ventricle.

venules, which are subsequently narrowed and occluded by intimal proliferation, fibrosis and organized thrombi. However, complete occlusion is uncommon. Hypertrophy in the walls of the pulmonary arteries may be quite striking (reactive prolifer-ative changes). The pulmonary capillary bed is generally unaffected. Thrombi in the pulmonary arteries are common(8). The gold standard for diagnosis of PVOD is biopsy, either during life or at autopsy. A diagnosis of PVOD should be suspected in a patient who develops dyspnea and fatigue with pulmonary venous hypertension, suggested by radiological evidence of interstitial haziness Kerley A, B and C lines and venous engorgement in lung (congested and edematous) if mitral valve-stenosis, cortriatrium, proximal pulmonary vein ostial stenosis, ASD, VSD and PDA have been excluded by echocardiography(9). Our case fulfilled these criteria.

Regardless of etiology, the disease is fatal in a majority of patients within two years of the onset of symptoms due to severe progressive pulmonary hypertension with right ventricular failure (l0). Management of this disease includes use of steroid, antimetabolites, anticoagulants (5) and vasodilator(11). In an earlier report, it was concluded that the pulmonary arterial vasodilating effect of nifedipine allowed additional time for veno-occlusive lesions to resolve before severe pulmonary arterial hypertensive changes occur (11). Recently, single lung transplant has been found to be effective (12).

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