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Case report

AN INTERESTING CASE OF COR PULMONALE

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ABSTRACT

A 14 yrs old boy presented with a history of breathlessness since 1 yr which had increased from 2 days, cough with expectoration and pedal edema since 7 days. In the past patient had history of kyphoscoliosis since birth and had a history of repeated URTI. On examination he had tachycardia, tachypnea, raised JVP, kyphoscoliosis, bilateral pitting edema. Respiratory auscultation revealed bilateral fine crepitations and wheezes. On investigation haemoglobin: 14.6, T.L.C: 20,000,chest X-ray: kyphoscoliosis with cardiomegaly. Clinical diagnosis of corpulmonale due to kyphoscoliosis was achieved and was confirmed with 2D echo. ECG showed RVH. The patient was treated with oxygen, diuretics, antibiotics, bronchodilators. Patient improved and was discharged on bronchodilators and was asymptomatic on follow up.

Key words: Corpulmonale, Kyphoscoliosis.

INTRODUCTION

Kyphoscoliosis is a disorder characterized by progressive deformity of Spine consisting of lateral and posterior curvature.In majority, it is of idiopathic etiology. Deformity results in shortening of height. Patients can be asymptomatic**.** Mobility of the chest wall is impaired, the chest wall is stiff and lung volumes are restricted. Hypoventilation can occur due to small tidal volumes and increased dead space ventilation. V/Q mismatch leads to significant hypoxia, and can progress to symptoms of Corpulmonale.[1]Certain persons with longstanding dorsal kyphoscoliosis develop pulmonary hypertension and corpulmonale. We had a young patient presenting with the same. Respiratory failure due to kyphoscoliosis, leading to corpulmonale, has rarely been reported, in spite of the fact that kyphoscoliosis is common. This fact led us to report this case.[2]

CASE

A 14 yrs boy presented with a history of breathlessness since 1 yr which was initially on exertion but had increased to breathlessness at rest since 2 days, cough with expectoration and pedal edema since 7 days. In the past patient had kyphoscoliosis since birth and had a history of repeated URTI.

On examination he had tachycardia (120 bpm), tachypnea (28/min), raised JVP (6cm), kyphoscoliosis, and bilateral pitting edema.

Respiratory ascultation revealed bilateral fine crepitations and polyphonic wheezes. The cardiac auscultation revealed loud P2 and there was also evidence of ejection systolic murmur in pulmonary area. On investigation haemoglobin: 14.6, T.L.C: 20,000/cmm, ABG was suggestive of type II respiratory failure (pH 7.37,pO_2 22mm Hg pCO2 52 mm Hg.,spo2:84%), 2D Echo showed severely dilated RA and RV , grade III TR, severe pulmonary hypertension 78mm Hg.

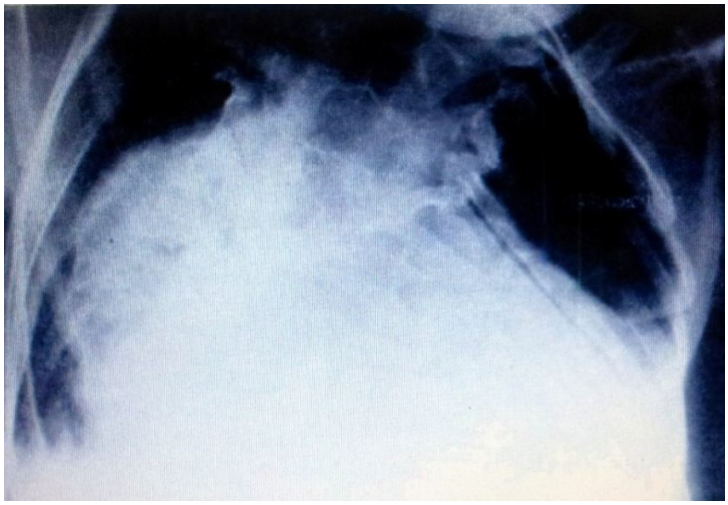

Fig:1. Chest X-ray showing Kyphoscoliosis with Cardiomegaly

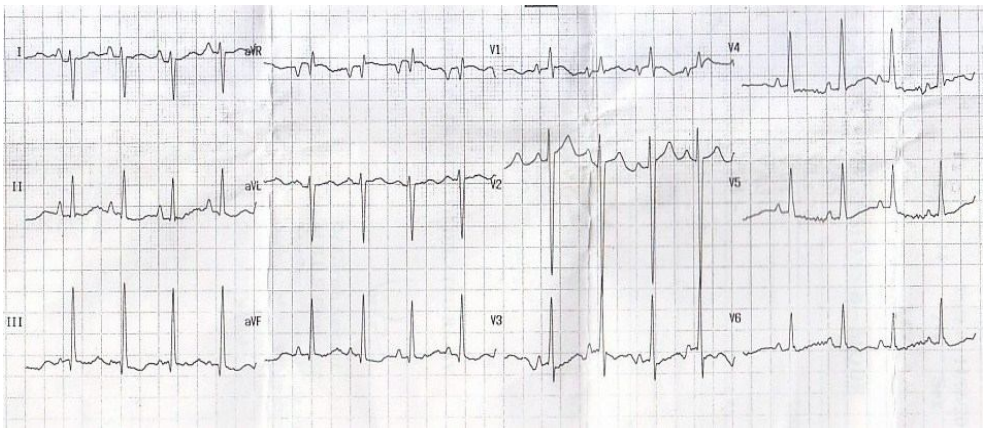

Fig: 2. ECG was showing 'P' Pulmonale, right axis deviation and right ventricular hypertrophy

Treatment

Controlled oxygen therapy, intermittent positive pressure respiration, digitalis, diuretics, antibiotic, bronchodilators (inhalational and intravenous).

The patient improved with treatment and was discharged on bronchodilators and diuretics.

Follow up

The patient is doing well with bronchodilators and diuretics.

DISCUSSION

Several pathophysiologic features contribute to respiratory dysfunction in kyphoscoliosis. Underlying problem is increased work of breathing resulting from poorly compliant chest wall. Distortion of chest wall causes under the ventilation of some regions of lungs, microatlectasis, ventilation perfusion mismatch and hypoxemia. [3] Common complication of kyphoscoliosis is pulmonary hypertension and corpulmonale. Hypoxemia and hypercapnia are important for the development of pulmonary hypertension. However increased resistance of pulmonary vessels results from compression especially in areas where chest wall is distorted. Exertional dyspnea is the most common symptom. PFT shows decreased TLC, VC and FRC. Chronic respiratory insufficiency and corpulmonale are the end results of severe kyphoscoliosis, the level of respiratory difficulty correlates with severity of chest wall deformity.[4]Cor pulmonale is associated with kyphoscoliosis when the external angle of the scoliosis is more than 100" and the kyphosis is more than 20°, with a vital capacity less than one liter[5].

In contrast to chronic pulmonary emphysema, the alveolar hypoventilation in kyphoscoliosis is due to anatomic striction of the chest rather than to bronchial obstruction and uneven distribution of inspired air. The factors responsible for the increased pulmonary resistance is believed to be mechanical compression of the vascular bed, anatomic thickening of the precapillary vessel wall, and effects of hypoxemia. [6]

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