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Pulmonary Assessment in Immune Mediated Connective Tissue Disorders

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ABSTRACT

Objective: Immune mediated connective tissue disorders (CTD) are known for their multisystem involvement. Pulmonary manifestations are obscured by more pronounced involvement of other systems. The search for pulmonary disease in these patients may bring to light mild to moderate and even asymptomatic involvement of respiratory system, thereby reducing the burden of pulmonary morbidity.

Material & Methods: This is an open labeled study of 25 consecutive outdoor patients with known CTD. They were evaluated for overt/obscure pulmonary disease by history taking, clinical examination, Chest X-ray & Spirometry, out of which selected patients were subjected to 2DECHO & HRCT thorax.

Results: Out of the total 25 patients, 7 were asymptomatic. In symptomatic patients, breathlessness on exertion was the predominant respiratory complaint. Large number of patients (60%) denied respiratory symptoms as their chief complaint, although they admitted being symptomatic on direct questioning. On Spirometry, 16 patients had mild to severe restrictive pattern, 5 had mixed obstructive-restrictive pattern. The remaining had a normal PFT. The predominant findings on HRCT were ground glass opacities, honey combing & pleural thickening.

Conclusion: Spirometry & HRCT thorax were abnormal in significant number of asymptomatic patients, thereby making them important investigations to detect early pulmonary disease & grade their severity.

INTRODUCTION:

Some diseases like collagen vascular diseases have no definite aetiology and have complex clinical patterns. They involve the whole of connective tissue system and originate from degeneration of collagen from ground substance, muscle, fibrin and other proteins. Most of them are insidious in onset and progress gradually to involve all systems of the body. They can be either controlled or symptomatically improved. They require persistent and lifelong monitoring, so it is important to detect these diseases in their early stage. In the present study patients with connective tissue disorders were examined clinically and investigations were carried out to detect early involvement of respiratory system.

MATERIALS AND METHODS:

The study is carried out on 25 patients with diagnosis of immune mediated connective tissue disorder (CTD) including SLE, Rheumatoid arthritis, Scleroderma, MCTD, Sjogren's syndrome, Polymyositis, Ankylosing spondylitis, vasculitis. All patients were evaluated for pulmonary disease and underwent X-ray chest, echocardiogram, PFT, HRCT in some cases.

OBSERVATION AND DISCUSSION:

Out of 25 patients, maximum patients were of SLE (40%), followed by RA (32%), scleroderma (16%), MCTD (12%). No patients of Polymyositis, Sjogren's syndrome, Ankylosing Spondylitis, vasculitis was there in my study.

Overall most common chest symptom was breathlessness on exertion (60%) followed by cough in 46%, without expectoration in 36% and with expectoration in 12%. The most common signs were clubbing (40%) and crackles (36%).

Out of all patients meeting diagnostic criteria of SLE, 40% were asymptomatic. Cough was the most common symptom in 50%, associated with dyspnea on exertion in 30% of patients. Out of all patients meeting diagnostic criteria of RA, 25% were asymptomatic. Cough and breathless on exertion were equally common in 62.5% of patients. Breathlessness on exertion was the most common symptom in MCTD and Scleroderma.

Table 1: Aetiology wise symptom distribution

Symptoms	SLE (n-10)	RA (n-8)	SC (n-4)	MCTD (n-3)	TOTAL (n-25)
Asymptomatic	4 (40%)	2 (25%)	1 (25%)	-	7 (28%)
Dyspnea	3 (30%)	5 (62.5%)	3 (75%)	3 (100%)	14 (56%)
Cough	5 (50%)	5 (62.5%)	1 (25%)	1 (33%)	12 (46%)
Chest pain	2 (20%)	1 (12.5%)	-	-	3 (12%)
Haemoptysis	-	1 (12.5%)	-	-	1 (4%)

Table 2: PFT Evaluation

PFT	SLE	RA	SC	MCTD	TOTAL
Normal	1	2	0	1	4 (16%)
Mild restriction	4 (40%)	1	2	0	7
Moderate restriction	2	1	2	2 (66%)	7
Severe restriction	1	1	0	0	2
Mixed	2	3 (37.5%)	0	0	5
Restriction only	7 (70%)	3 (37.5%)	4 (100%)	2 (66%)	16 (64%)

40% of patients had normal chest X-rays. 50% patients of SLE and 37.5% of patients with RA had normal chest X-rays. In SLE commonest X-ray chest abnormality was blunting of CP angle and reticular pattern in 30% cases. In RA honeycombing was most common in 37.5% patients followed by reticular and nodular pattern. In scleroderma reticular pattern was the most common and reticular and honeycombing in MCTD.

While doing PFT, only 1 patient (10%) with SLE with or without symptoms had normal PFT while 25% patients with RA had normal PFT. Commonest result of spirometry was restriction pattern which was seen in 64% cases. Restrictive pattern was more commonly present in SLE and scleroderma. Obstructive and restrictive pattern was more found in patients with RA.

HRCT was done in selected cases that had significant abnormalities on PFT. None of the HRCT was normal. In RA, 37.5% patients had ILD and Bronchiectasis and 12.5% of patients had bronchiolitis obliterans organising pneumonia. In scleroderma, ILD was prevalent in 50% of cases. 20% of patients with SLE had pleurisy, 20% had ILD and 10% had Bronchiectasis.

SUMMARY:

Among the 25 cases of CTD, 40% were of SLE, 32% were of RA, 16% of scleroderma and 12% of MCTD. Common age group were 31-40 years (36%) followed by 41-50 years (24%). Female to male ration in this study was 9:1 for SLE and 7:1 for RA.

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Out of all, 28% patients had no respiratory symptoms. While most common symptoms were breathlessness on exertion (60%) and cough (48%). Common signs were clubbing (40%) and crackles (36%). Most common findings on chest x-ray were reticular pattern (32%) and honey combing (24%). In HRCT thorax ground glass opacity was the most common pattern (60%). Pleural thickening was also present in 60% of cases. Honey combing was present in 40% of cases. Most of the patients undergoing HRCT had advanced lung disease. Pleural thickening was more common in patients with SLE, ground glass opacity and honey combing was more commonly found in patients with RA.

Spirometry was normal only in 16% of patients. Restriction pattern was the commonest finding in 64% cases. 20% patients had mixed obstructive and restrictive pattern. 40% patients had no respiratory symptoms but 75% of them were having mild restrictive pattern on spirometry. Restrictive pattern was more commonly present in SLE and scleroderma. Obstructive and restrictive pattern was more found in patients with RA.

Echocardiography showed that 56% patients had no pulmonary arterial hypertension. 40% of patients with SLE had pulmonary hypertension, while 50% of patients with RA and scleroderma had some form of pulmonary hypertension. Overall 30% patients with SLE had pleural manifestations in form of pleuritis and pleural effusion. 30% of patients with RA and 50% of patients with scleroderma were diagnosed of having interstitial lung disease (ILD). Bronchiectasis was present in 37.5% of patients with RA.

CONCLUSION:

Breathlessness on exertion and cough were two most important pulmonary symptoms in these patients. x-ray chest is insensitive mean for diagnosing severity of pulmonary disease in immune mediated connective tissue disorders while spirometry and HRCT are important means to detect early pulmonary involvement in these disorders. Spirometry was abnormal in 80% of asymptomatic patients. Present study highlights that most of the symptomatic patients presented with extensive involvement of lung parenchyma as well as with poor lung function. Most of the asymptomatic patients who presented at earlier stage had mild lung function abnormality. Periodic respiratory assessment and PFT can be extremely useful means to detect subclinical involvement in all connective tissue disorders.

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