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Research Article

PULMONARY MANIFESTATIONS OF SJOGREN'S SYNDROME-A STUDY ON 42 PATIENTS

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ABSTRACT

Introduction: Sjogren's syndrome is defined by an autoimmune condition where there is influx of WBCs in the exocrine gland leading to dry mouth, dry nasal cavity and dental erosion. Sjogren's syndrome can affect musculoskeletal, gastrointestinal, neurological, renal, cardiovascular and pulmonary systems. On extensive chart review we found that there are not many studies documenting the findings of CT scan of lungs in patients with Sjogren syndrome. Hence this study serves that purpose. **Methods:** We selected 42 patients who presented to the outpatient office between January 2017 to December 2021 who had diagnoses of Sjogren's syndrome and had CT scan lung performed for various diagnostic purposes. We analyzed the CT scan findings for the most common pulmonary changes seen in patients with Sjogren's syndrome. **Results:** Our sample included 42 patients out of which 34 were female and 8 were male with mean age of femalesbeing 54 (age range-42-68) and of males being 60(age range-45 to 72). Out of the 30 patients who had changes on CT scan, 12 patients had bronchiolar abnormalities, 5 patients had Interstitial lung disease, 3 had bilateral interstitial infiltrates, 3 had ground-glass opacities with subpleural and basilar predominance, 3 had Reticular abnormalities, with or without traction bronchiectasis, 2 had Honeycombing, 1 had peripheral parenchymal consolidations with air bronchograms and variable associated ground-glass opacities, and 1 had Cystic lung disease. **Conclusion:** Our study concluded that CT scan are a superior imaging modality in characterizing the disease progression in the lungs in patients of Sjogren's syndrome. Chest X-ray is useful to detect only advanced imaging findings like pleural effusion or infiltrates. These findings are rare and would not be seen commonly.

Keywords: Sjogren's syndrome, pulmonary, lung, CT scan.

INTRODUCTION

Sjogren's syndrome is defined by an autoimmune condition where there is influx of WBCs in the exocrine gland leading to dry mouth, dry nasal cavity and dental erosion. It affects the eyes, mouth, nose, pharynx, larynx and vagina.[1] Sjogren's is further classified into primary disorder or secondary disorder which has another underlying autoimmune disorder.[2] Sjogren's syndrome can affect musculoskeletal, gastrointestinal, neurological, renal, cardiovascular and pulmonary systems. The symptoms like dry eye and dry mouth are present in more than 90 percent of the patients.[3] There is no universally accepted diagnostic criteria for Sjogren syndrome but diagnosis would be suspected in patients who have sicca symptoms with more than 1 of the following-positive salivary gland biopsy with evidence of chronic inflammatory infiltrate an exuberant gland, positive blood test for antibodies to anti SSA or anti SSB ENT gin, nonspecific indications assistant manifestation.[4] Initial test for evaluation of sicca symptoms include schirmer test, corneal staining with current and an stimulated salivary fluid collection.[5] On extensive chart review we found that there are not many studies documenting the findings of CT scan of lungs in patients with Sjogren syndrome. Hence this study serves that purpose.

MATERIAL AND METHODS

We selected 42 patients who presented to the outpatient office between January 2017 to December 2021who had diagnoses of Sjogren's syndrome and had CT scan lung performed for various diagnostic purposes. We collected data from two rheumatology clinic which serves 20,000 patients per year. All patients that were selected were nonsmokers, defined as no history of smoking since the past 15 years. Since there is no universally accepted definition for Sjogren syndrome, we used the following definition for diagnosing patients with Sjogren's:

- patients with sicca symptoms and ≥ 1 of the following:
- positive blood test for antibodies to anti-SS-A or anti-SS-B antigen
- positive salivary gland biopsy with evidence of chronic inflammatory infiltrate in exocrine glands
- nonspecific and/or organ-specific symptoms indicative of systemic manifestations

Informed consent was obtained from each patient for inclusion in the study. Permission was obtained from the institutional ethics committee. CAT scan specification were as follows: Scanner type-multi detector helical (spiral) detector rows ≥ 4, kV-100 to 140 acceptable for standard sized patient, mAs-set in combination with kVp to meet CTDIvol specifications, Max. Tube Rotation Time-≤ 0.5 seconds, Pitch (IEC Definition)-Between 0.7 and 1.5, Respiration-single breath hold full inspiration, Reconstructed image width (nominal width of reconstructed image along zaxis) ≤ 1 mm, CTDIvol-≤ 3 mGy for standard size patient, Anatomical Coverage-Lung apex through the lung bases, Gantry tilt-None, Display FOV-1 cm beyond the rib cage, Display window width / level-Lung: Allow adequate visualization of the lung parenchyma and intraparenchymal airways and vessels. WW = 1200 – 1500 HU WL = -550 – - 700 HU Media stinum: Allow visualization of the mediastinal and hilar vessels and allow distinction of the chest wall musculature from subcutaneous fat. WW = 250 – 450 HU WL = 40 – 80 HU, Additional reformats-MPR-Multiplanar reformation; MIP-Maximum intensity projection.

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RESULTS

Our sample included 42 patients out of which 34 were female and 8 were male with mean age of females being 54 (age range-42-68) and of males being 60 (age range-45 to 72). All 42 patients have had some kind of respiratory symptoms at the time of the CT scan. 14 patients complained of chronic cough, 20 patients have complained of dyspnea on exertion, 8 patients complained of dyspnea at rest. The mean age of onset in men was 24 years (age range- 8- 32 years) and in female was 11 years (age range-2 to 19 years). The median age of respiratory symptom onset for men was 58 years (age range 45-67 years) and for female was 56 years (age range- 43-65 years). Thirty (72 percent) patient were found to have abnormal CT scan findings. Out of the 30 patients, 12 patients had bronchiolar abnormalities, 5 patients had Interstitial lung disease, 3 had bilateral interstitial infiltrates,3 had ground-glass opacities with subpleural and basilar predominance, 3 had Reticular abnormalities, with or without traction bronchiectasis, 2 had Honeycombing, 1 had peripheral parenchymal consolidations with air bronchograms and variable associated ground-glass opacities, and 1 had Cystic lung disease. Of those patients who were found to have bronchiolar abnormalities, 7 had bronchiectasis, 3 had bronchiolar wall thickening and 2 had both bronchiectasis and bronchiolar wall thickening. We found that out of 30patients who had pulmonary radiological findings, 75 percent of the patients had lower lung field involvement. Bilateral infiltrates, ground glass opacities and peripheral parenchymal consolidation were more common in lower lung fields. Micro nodules were also appreciated in 20 percent of the patients. Majority had middle lung involvement. One patient had parenchymal cystic disease and the cysts were equally distributed in the bilateral lungs in all lung field. We did not find pleural effusion in any of our patients.

DISCUSSION

Many systemic autoimmune disease have shown to have pulmonary manifestation. Rheumatoid arthritis and Systemic Sclerosis were the most common systemic autoimmune disease to have pulmonary manifestation.[6] In Sjogren's syndrome the physiology behind pulmonary involvement is unclear. Sjogren's syndrome is characterized by invasion and then by destruction of the exocrine glands by the WBCs.[7] The initial symptoms of Sjogren's syndrome involved the upper respiratory tract and will manifest with dry mouth and dry nose.[8] Clinical signs are not enough to estimate be extent of pulmonary involvement and radiological findings are necessary. Many studies have reported that Sjogren syndrome primarily involves the bronchus and manifest as bronchiectasis or bronchial wall thickening.[9-10] This was similar to the findings found in our study. Parenchymal involvement by inflammation and scarring of the pulmonary tissue is also very common in Sjogren syndrome.[11] The purpose of our study is to evaluate the most common pulmonary radiological findings in patient with Sjogren's syndrome. To avoid any changes in the lung from secondary autoimmune condition or smoking only patients with no smoking history and who had no diagnosed secondary autoimmune condition were selected. This selection maximized the possibility that the abnormal findings seen on CT scans were indeed caused by primary Sjogren's syndrome and did not represent sequelae of other pathologic agents that affect the lung. Many studies have done testing through pulmonary function test in patient with Sjogren's syndrome and they found that the diffusion lung capacity of carbon monoxide was significantly reduced in patients of Sjogren's syndrome likely secondary to scarring.[12-13] Out of the 30 patients, 12 patients had bronchiolar abnormalities, 5 patients had Interstitial lung disease, 3 had bilateral interstitial infiltrates, 3 had ground-glass opacities with sub pleural and basilar predominance, 3 had Reticular abnormalities, with or without traction bronchiectasis, 2 had Honeycombing, 1 had peripheral parenchymal consolidations with air bronchograms and variable associated ground-glass opacities, and 1 had Cystic lung disease. Chest X ray is not considered the best imaging modality in detecting early stage pulmonary radiological findings in patients of Sjogren's syndrome.[14] Many studies described radiological imaging findings in chest radiograph in patient with Sjogren's syndrome and they found that the most common finding was changes in the interstitium, linear reticulation and pleural effusion. Pleural effusion is a very rare finding in Sjogren's syndrome and is a radiological findings seen in advanced Sjogren syndrome.[15] Few cases of primary Sjogren's syndrome with pleural effusion have been reported. Indeed, only 13 cases of primary SS with pleural effusion have been described so far.[16-18] The treatment of patients with pleural effusion in Sjogren's syndrome is not certain as the number of cases documented so far has been very low. A general consensus says that suppressing the systemic inflammatory response helps in decreasing the pleural effusion in some patients.[19] Hence chest x-rays are not a modailty of choice in patients with early stage Sjogren's syndrome as they are not able to detect subtle pulmonary changes as compared to HRCT. Modalities to detect earlier pulmonary involvement in patients with Sjogren's disease are bronchioloalveolar lavage, lung biopsy and pulmonary function testing.[20] In our study we found that bronchial involvement was the most common radiological finding in Sjogren's syndrome and this is in line with the other similar studies come to the scene the conclusion.[10] As a matter of fact patients with rheumatoid arthritis and systemic lupus erythematous have also been found to have more bronchiolar involvement as compared to parenchymal involvement.[21] Majority of our patients had respiratory symptoms and mainly presented with cough and dyspnea. The airway disease in Sjogren's syndrome is most commonly involved lung area which manifest mainly by coughing. The airway disease has been associated with Sjogren's disease primary pathophysiology of destruction of the endocrine glands. One study demonstrated that even if patient does not have any symptoms the histopathology of the bronchial wall showed extra glandular cell infiltration.[22] One study reported that the damage to be the central and the peripheral bronchi and bronchioles through cellular infiltration causes bronchiectasis.[23] Some studies have performed pulmonary function testing in patients who had radiological findings of bronchiectasis and parenchymal disease and they found that there was no significant changes in the pulmonary Function Test in majority of the patients.[24-25] Our study concluded that CT scan are a superior imaging modality in characterizing the disease progression in the lungs in patients of Sjogren's syndrome. Chest X-ray is useful to detect only advanced imaging findings like pleural effusion or infiltrates. These findings are rare and would not be seen commonly.

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