Squamous Cell Carcinoma Lung with Progressive Systemic Sclerosis

Susmita Kundu, Ritabrata Mitra, Swapnendu Misra, Sumit Chatterjee

Abstract

Association between progressive systemic sclerosis (PSS) and lung malignancy is rare yet well recognised. In order of frequency bronchioloalveolar carcinoma (BAC) is most common followed by squamous cell carcinoma, oat cell carcinoma, anaplastic carcinoma of lung. We present a 55 year old non-smoker male patient with PSS, who presented to us with progressively increasing shortness of breath and dry cough. Initially clinico-radiologically he was provisionally diagnosed as a case of non-resolving pneumonia. Subsequent investigations revealed it is a case of squamous cell carcinoma of lung with PSS.

Introduction

Hale and Schatzki suggested a possible relationship between PSS and lung malignancy in 1944. The relative risk of lung malignancy in patients with PSS has been placed at up to 16.5 times that of normal persons. Bronchioloalveolar carcinoma which accounts for less than 5% of all lung cancers is the most common histological type associated with PSS, followed by Squamous cell carcinoma, oat cell carcinoma, anaplastic carcinoma and mesothelioma. The exact incidence of squamous cell carcinoma in PSS is unknown. The strong relationship between lung malignancy and PSS may be due to scarring of pulmonary tissue. The so-called scar cancer is believed to be caused by transformation of hyperplastic epithelium to metaplasia and finally to neoplasia under the conditions of chronic inflammation with some unknown etiological factors.

Case Report

A 55yr old male non smoker patient was admitted to our department with progressively increasing dyspnoea and dry cough for last 4 months. He had a 4 year history of progressively increasing tightening of skin involving the whole body but without any history of Raynauds’ phenomenon. On admission, he looked tachypneic. He had a masked facies (Figure 1) and skin of his face and all four limbs were thickened. There were areas of hyperpigmentation over abdomen and right knee joint and hypopigmentation over upper forehead and both pinna. Clubbing was present. Digital infarcts were present over middle and index fingers of both hands. Examination of the Respiratory system was unremarkable except for the presence of end inspiratory velcro type crepitations involving both infra scapular regions. Heart sounds were normal. Examination of other systems did not detect any abnormality.

In laboratory findings, Complete blood count, urinalysis and blood biochemistry were normal. ANA was positive in more than 1:640 titer and it was fine speckled with nucleolar pattern. RA factor was negative, serum ACE was within normal limit. Anti scl70 antibody was strongly positive.

Chest X ray (Figure 2) shows bilateral inhomogenous opacities involving both lower zones. HRCT Thorax (Figure 3) shows bilateral focal areas of sub segmental consolidation, bilateral minimal pleural effusion with thickening.

Pulmonary function test showed restrictive defect. Upper GI Endoscopy revealed hiatus hernia. Echocardiography revealed no evidence of pulmonary hypertension.

Fibreoptic bronchoscopy revealed diffuse luminal narrowing with thickened mucosa and no endobronchial mass was found. BAL fluid cultures (Routine, AFB and Fungal Culture) revealed no growth.

Papanicolaou stain of BAL fluid revealed occasional dysplastic cells.

Bronchial brush cytopathology showed discrete and clusters of neoplastic epithelial cells with large hyperchromatic nuclei (black arrow) suggestive of Squamous cell carcinoma (Figure 4).

Discussion

PSS is a multisystem disease characterised by varying degrees of vasculitis, fibrosis and inflammation of skin and internal...
Although dermatological manifestations dominate the clinical picture of this multisystem disease, it is the visceral organ involvement that determines survival. Interstitial fibrosis is almost always present in scleroderma lung.\(^5\) Less commonly cystic lesions and pleural effusion can be noted.

The concept of lung malignancy arising in the vicinity of pulmonary scars formed previously from tuberculosis, infarction, abscesses or interstitial fibrosis etc. has been well established by a number of studies.\(^6\) The scar concept is also applied to the relationship between lung malignancy and PSS, although the basic process is not yet clarified.

In scleroderma lung, the atypical proliferation of alveolar epithelium occurs near fibrotic lung tissue. Fibrotic lung tissue blocks lymphatic channels. As a result, there is accumulation of carcinogens. Accumulated carcinogens in presence of a long standing immunological imbalance initiates a pathological process by which metaplastic alveolar epithelial cells ultimately turn neoplastic.\(^7\)

Our case was a 55 year old non-smoker male with PSS. PSS is uncommon in males. The final diagnosis also surprised us as squamous cell carcinoma is very rare in PSS.

**References**