

Total Pulmonary Fibrosis with Compensatory Emphysema

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Abstract

We report a case of a 60-year old male, previously treated for pulmonary tuberculosis, who presented with complaints of bilateral mass descending per anterior nares and nasal block. Imaging confirmed the diagnosis of bilateral nasal polyposis and additionally revealed total pulmonary fibrosis of left lung, elevation of the left hemi-diaphragm, herniation of the right lung, and reduced heart borders. A diagnosis of bilateral nasal polyposis and total pulmonary fibrosis of the left lung as sequelae of pulmonary tuberculosis with compensatory emphysema of the right lung was made. What was interesting was that despite the totally fibrosed lung, the patient’s SpO₂ was normal, showing the degree of compensation by the right lung and accounting for why the total pulmonary fibrosis was only incidentally diagnosed. Pulmonary fibrosis is an irreversible complication with permanent respiratory function compromise - as there is no cure, slowing down the progression is the main form of treatment with pneumectomy. We present this case to highlight the fact that even though compensatory emphysema of one lung could allow for normal oxygenation and prevent complete respiratory compromise, despite total lung fibrosis of the other, the sequelae of tuberculosis and their damaging effects, especially in the elderly, need more emphasis. We also briefly discuss the need for better understanding the immunopathogenesis of pulmonary impairment after TB (PIAT).

Keywords: compensatory emphysema, nasal polyposis, pulmonary tuberculosis, total pulmonary fibrosis

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Introduction

Tuberculosis (TB) in older adults is expected to present major challenges to global tuberculosis control in the future.¹ As destructive as tuberculosis itself is, its sequelae are equally damaging; Post tuberculosis sequelae include pulmonary fibrosis, bronchiectasis, cavitation, etc.² Pulmonary fibrosis is the replacement of normal lung parenchyma with collagenous tissue, which results in thickening and stiffening of the lung walls.² Compensatory emphysema, considered a misnomer, refers to the overinflation or increase in the size and function of one lung when the other is destroyed or temporarily

useless.^{2,3} Despite the relatively high prevalence of Post-TB lung dysfunction and the fact that patients with pulmonary impairment after TB (PIAT) have a reduced quality of life, the specific host and pathogen factors that are responsible for the impairment are largely unclear and it could go unrecognized in many cases.²

Case presentation

A 60-year old male patient, who was a chronic smoker and who was previously treated for pulmonary tuberculosis, presented to the ENT Out-

Patient Department with complaints of bilateral nasal mass descending per anterior nares and nasal block. General examination revealed that the patient was dyspnoeic and anemic. There was no clubbing, pedal edema, palpable lymph nodes, jaundice or cyanosis. Routine blood investigations were normal except for the Haemoglobin being 9.5gm%. On examination of the respiratory system, breath sounds were found to be absent over the left lung, but normal vesicular breath sounds were auscultated on the right side. Decreased chest expansion was also observed. The bilateral polyps were visible during an ENT examination. CT scan confirmed the bilateral nasal polyposis. Additionally, the X-Ray Chest revealed total pulmonary fibrosis of the left lung, elevation of the left hemi-diaphragm, herniation of the right lung, and reduced heart borders (Figure 1). Chilaiditi's sign which is the incidental radiographic finding of the bowel positioned between the right diaphragm and the liver was also observed.

Figure 1: X-Ray Chest revealing the total pulmonary fibrosis (left)



The patient was diagnosed as bilateral nasal polyposis with total pulmonary fibrosis of the left lung and compensatory emphysema of the right lung as sequelae of pulmonary tuberculosis. Interestingly, in spite of a totally fibrosed lung, the patient's SpO₂ was found to be normal (100%) indicating the degree of compensation by the contralateral lung which effectively maintained normal oxygenation levels. This could have accounted for why the diagnosis of the totally fibrosed lung with compensatory emphysema of the other lung was only incidentally made when the patient visited the hospital for a nasal problem. The patient underwent a biopsy of the right nasal mass which was subsequently

reported as squamous cell carcinoma. The patient will be reassessed during subsequent follow-up visits and further management will be planned.

Discussion

Pulmonary fibrosis causes a restrictive ventilatory defect that is irreversible with permanent respiratory function compromise.^{2,4-7} Recent research identifies that cytokines are involved in the development of fibrosis.^{2,4,5} The activation of TGF- β pathway is associated with increased collagen levels.⁴ This causes excessive collagen deposition and fibrotic scarring.⁴ Studies point that an imbalance in interleukins (IL) IL-1RA and IL-1 β may also plausibly contribute towards fibrosis.^{2,5} Studies suggest the need for further investigation in the area of fibrinogenic cytokines as TGF- α , TGF- β and IL-1 β , collectively, could plausibly contribute towards the restrictive ventilatory defects seen in patients with a history of tuberculosis.²

Respiratory function compromise may be observed as a decrease in forced vital capacity (FVC) and an increase in FEV₁/FVC ratio (FEV₁=Forced expiratory volume₁).⁶ In the patient we have described, the occurrence of compensatory emphysema prevented total respiratory compromise and allowed for normal oxygenation levels. In this patient, the entire left lung was fibrosed. Generally, the incidence of unilateral pulmonary fibrosis is more common on the left side (63%).⁷ Recent studies are in concordance with the more common occurrence of unilateral left lung fibrosis, the left lung being more affected due to the anatomy of the left bronchus (longer and 15% narrower).⁷ It's more horizontal course affects the drainage of secretions, which results in increased oxygen tension.⁷ The reduced oxygen favors the progression of fibrosis.⁷

With no cure, treatment focuses on delaying disease progression and pneumectomy.⁸ Pneumectomy is generally suggested for treating a lung that is destroyed by tuberculosis as research shows that pulmonary fibrosis, post tuberculosis, is associated with an increased risk of lung cancer in the form of scar carcinoma.^{9,10} Prevention is possible if there is early treatment and diagnosis.⁶ Disappointingly, however, as was the case in this patient, the sequelae of tuberculosis often go unrecognized and are only discovered much later.⁶

However, there is a felt need for more translational studies that would help better understand the immunopathogenesis of pulmonary impairment after TB (PIAT); this understanding would then help choose appropriate prophylactic and therapeutic strategies.² Although specific host and pathogen factors responsible for lung impairment are largely unclear, given that excessive inflammation and elevated expression of lung matrix-degrading proteases are seen in tuberculosis, it could be probable that host immune responses play a dominant role in lung damage.² To that extent, as Ravimohan *et al.* point out, a hypothesis that is largely untested but plausible is that variability in host genes that modulate these immune responses could account for how severe the lung impairment is.² The pathogenesis of post-TB lung impairment needs to be looked at from the perspective of dysregulated immune responses and immunogenetics and the management of TB could be pulmonary impairment specific and could target specific underlying immunopathological mechanisms.² Therefore, we could see in the future, drugs that harness the host's immune response being administered in addition to antitubercular treatment, the therapies being specifically tailored to target specific immune pathways based on the patient's inflammatory and immunogenetic profile.²

Conclusion

We present this case report of total pulmonary fibrosis with compensatory emphysema to highlight how even total pulmonary fibrosis may go unnoticed or be found only incidentally when compensatory emphysema occurs additionally. We emphasize that there is a need more emphasis to be given to the sequelae of tuberculosis and their damaging effects, especially in the elderly. We briefly address the need to better understand the immunopathogenesis of pulmonary impairment after TB (PIAT) and start using that knowledge for better prevention and treatment.

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Conflicts of interest: Nil

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