Case Report

Bronchocentric granulomatosis with extensive cystic lung disease in tuberculosis: An unusual presentation

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ABSTRACT

Tuberculosis is known to cause both cystic lung disease and bronchocentric granulomatosis (BCG). However, both are rare manifestations of this common disease. We report a case of BCG with extensive cystic lung disease in a young female who presented with fever, weight loss, and recurrent pneumothoraces with respiratory failure. Early diagnosis and treatment are imperative, as appropriate therapy may be life-saving in such cases.

KEY WORDS: Bronchocentric granulomatosis, cystic lung disease, tuberculosis

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INTRODUCTION

Bronchocentric granulomatosis (BCG) is a rare, often destructive, granulomatous lesion of the bronchi and bronchioles.[1] Severe cystic lung disease secondary to pulmonary tuberculosis (TB) is also not commonly reported, though fibrocystic lung lesions are seen commonly.[2] The coexistence of these two conditions together in a single patient has not been reported. Here, we present this unusual association in a young female, where the diagnosis was confirmed by surgical lung biopsy.

CASE REPORT

A 14-year-old, previously, healthy female presented with the complaints of fever, since 3 months followed by shortness of breath and weight loss since 1 month. There was a positive history of pulmonary TB in the mother, who was currently on treatment. The father of the patient was a laboratory technician at the local DOTS center. A chest X-ray done outside showed bilateral, randomly distributed nodules. A diagnosis of miliary TB was made, and she was started on anti-tubercular therapy (ATT) with 4 drugs (isoniazid, rifampicin, pyrazinamide, and ethambutol). Two sputum examinations for acid-fast *Bacilli* (AFB) were negative. Two weeks after starting ATT, she developed worsening of her symptoms, and a repeat X-ray chest showed bilateral pneumothoraces. Bilateral intercostal chest tube insertion was done, and she was referred to our center for further management. On presentation to us, the patient was in severe distress, her pulse rate was 120/min, blood pressure was 90/60 mmHg, and respiratory rate was 36/min. On auscultation, chest revealed bilateral crackles and absent breath sounds over most of the hemithoraces. The patient was febrile (102 F) and was very thinly built. Arterial blood gas analysis on 60% FiO_2 venturi mask showed pH - 7.45, PaO_2-77 mmHg, $PaCO_2$-50 mmHg, HCO_3-34.5 mmol/L, and SpO_2-92%. Chest X-ray showed bilateral residual pneumothoraces with underlying collapsed lung with bilateral Intercostal chest tube drainage (ICD) tube *in situ*. Laboratory investigations revealed hemoglobin of 8 mg/dl, total leukocyte count 12,900 cumm with neutrophils of 80%. ESR levels were 58 mm/h. Serum urea was 18 mg/dl, and creatinine was 1.2 mg/dl. Serum sodium was 133 mEq/L, and serum potassium was 4.2 mEq/L. Total bilirubin was 0.6 mg/

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dL, direct bilirubin was 0.2mg/dL, aspartate aminotransferase was 32 U/L, and alanine aminotransferase was 38 U/L. Blood investigations did not reveal any eosinophilia. Serum IgE levels were normal (18 IU/ml). P-antineutrophilic cytoplasmic antibodies (P-ANCA) and C-ANCA were negative. High resolution computed tomography (CT) thorax revealed bilateral pneumothoraces with septae, multiple extensive cysts of varying sizes, and minimal ground glass haze in both the lungs [Figure 1].

In an attempt to drain the pneumothoraces better, two more intercostals chest drains were inserted under CT guidance; one on each side and attached to negative suction. However, the lungs still did not expand, and negative suction was tried but failed too. In order to establish a diagnosis and as an attempt to do some adhesiolysis to expand the lung, it was decided to do a video-assisted thoracoscopic surgery guided surgery on the left side. Intraoperatively, the lung was found to be collapsed with cysts of varying sizes visible over the lung surface and multiple thick pleural strands and adhesions were seen [Figure 2].

Intraoperatively, fiber optic bronchoscopy was performed through the endotracheal tube, and bronchial washings were taken. Bronchial washings were smear positive for *Mycobacterium tuberculosis*. GeneXpert test was performed on the bronchial washings, which was positive for *M. tuberculosis* and no rifampicin resistance was detected. Conventional TB culture was sent, which subsequently grew *M. tuberculosis*, sensitive to first-line ATT.

Histopathological examination of the surgical lung biopsy showed areas with extensive necrosis and cystic changes. The biopsy specimen tested positive for AFB. Silver staining was negative for fungal elements, and there was no evidence of vasculitis. There were areas in the sample which revealed extensive peribronchiolar necrotizing inflammation, with bronchial wall destruction with palisading granulomas, replete with giant cells and a few eosinophils. This histopathological picture was consistent with BCG [Figure 3].

The patient was continued on first line ATT and steroids (4 mg of dexamethasone, twice daily) were added. The fever and respiratory failure, however, did not respond. The patient was weaned of the ventilator on the 4th postoperative day, and one ICD tube was removed from both the sides and the oxygenation improved marginally. On the 10th postoperative day, the patient again developed a right-sided pneumothorax, which was confirmed by a bedside ultrasound examination. An ICD was re-inserted anteriorly. However, she sustained a cardiac arrest the same day and could not be revived.

DISCUSSION

BCG is a rare, destructive, granulomatous lesion of the bronchi and bronchioles that is generally believed to represent a nonspecific response to various types of airway injuries.[1] Around half of these cases are associated with asthma and allergic bronchopulmonary aspergillosis and may represent a histopathologic manifestation of fungal hypersensitivity among these patients.[3] Other

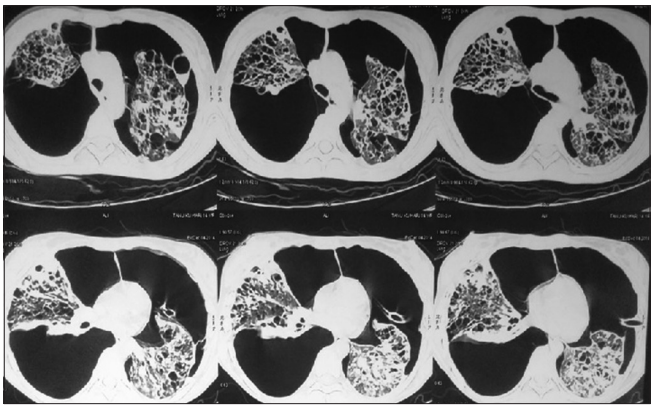

Figure 1: Computed tomography chest showing extensive bilateral pneumothoraces with multiple parenchymal lung cysts

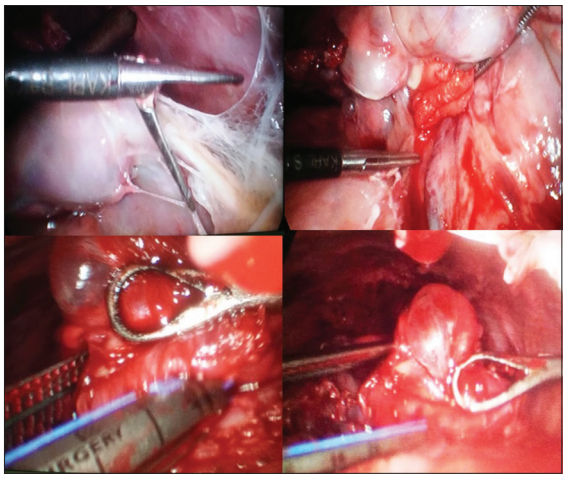

Figure 2: Intraoperative video-assisted thoracoscopic surgery picture, demonstrating multiple lung cysts of varying sizes

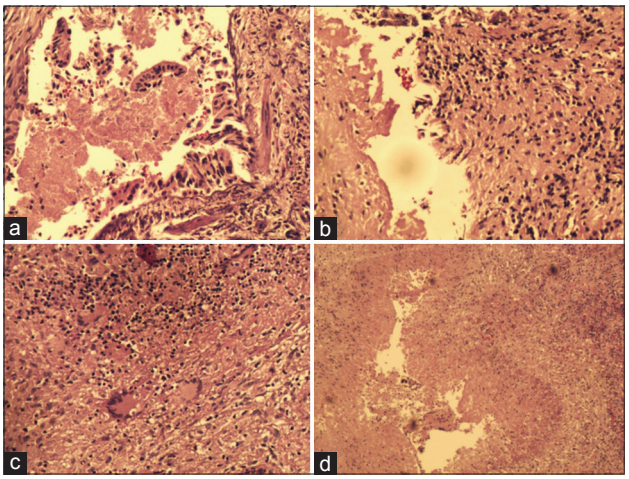

Figure 3: (a-d) Histopathology panel consistent with bronchocentric granulomatosis – extensive peribronchiolar necrotizing inflammation with bronchial wall destruction, pallisading granulomas replete with giant cells

reported associations include mycobacterial and fungal infections, rheumatologic diseases, granulomatosis with polyangiitis (Wegener's), diabetes insipidus, red cell aplasia, pulmonary *Echinococcosis*, bronchogenic carcinoma, and influenza infection.[4]

The presence of this lesion should generally be considered a nonspecific manifestation of lung injury and not an etiologic diagnosis. *M. tuberculosis* infection if implicated as a cause of lung injury, representing BCG should be treated at the earliest, to halt the lung injury. BCG is characterized by peribronchial and peribronchiolar necrotizing granulomatous inflammation. Bronchioles are more uniformly involved than the larger conducting airway. Subsequent destruction of airway walls and adjacent parenchyma lead to granulomatous replacement of mucosa and submucosa by palisading, epithelioid, and multinucleated histiocytes.[1,2,5] The radiographic presentation of BCG is varied, and pulmonary nodules, consolidation, mass lesions, etc., have all been reported.[6,7]

In our patient, the picture was also confounded by the presence of multiple lung cysts of varying sizes, which led to recurrent pneumothoraces and clinical worsening in our patient. The formation of lung cysts due to TB can be attributed to number of causes.

Caseating necrosis of the bronchial walls leading to cystic bronchiectasis as a distal extension of fulminant tubercular bronchitis and granulomatous involvement of the bronchioles may lead to a check-valve mechanism leading to cyst formation.[2]

TB produces an exudative, necrotizing inflammation in the lung. The phenotype and the final clinical picture, of such inflammation, are variable. Our patient had areas of extensive cystic damage bilaterally. However, there were multiple areas with histopathology pathognomic of BCG. Whether the BCG pattern is secondary to TB or a *de novo* entity is a matter of debate.[4] TB is a disease of antiquity, but its varied presentations and features never cease to surprise us.

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Conflicts of interest
There are no conflicts of interest.

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