

A Rare Cause of Pneumonia: Tracheobronchopathia Osteochondroplastica

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ABSTRACT

Tracheobronchopathia osteochondroplastica is a rare disorder with a benign course. It is characterized by the accumulation of diffuse cartilaginous and osseous nodules protruding into the mucosa of the trachea and bronchus. We report the case of a 50 year old male who presented with fever, cough and shortness of breath and he was diagnosed on bronchoscopy, radiologically and clinicopathologically to have Tracheobronchopathia osteochondroplastica.

Keywords: Tracheobronchopathia osteochondroplastica, bronchoscopy, radiology

INTRODUCTION

Tracheobronchopathia osteochondroplastica is a rare disorder with an unknown etiology. It is characterized by the accumulation of diffuse cartilaginous and osseous nodules protruding into the mucosa of the trachea and bronchus. TPO was first described in detail by Rokitsky and Wilms in 1857 after an autopsy.

We report the case of a 50-year old male who presented with fever, cough and shortness of breath and he was diagnosed on bronchoscopy, radiologically and

clinicopathologically to have Tracheobronchopathia osteochondroplastica.

CASE REPORT

A 50 years old male, resident of Mancherial district, Telangana who is a labourer by occupation presented with chief complaints of fever since 7 days, cough and shortness of breath since 5 days. Fever was high grade in nature, intermittent type, associated with chills. Cough was insidious in onset, productive in nature, but not associated with diurnal or postural variation. Cough was



Figure 1: Chest radiograph (posteroanterior view) showed a large non homogenous opacity with cavities on the right upper lobe.



Figure 2: Chest radiograph (posteroanterior view) taken after 5 days showing worsening.



Figure 3: Bronchoscopy revealed multiple nodules protruding into the lumen of trachea from the antero lateral wall.

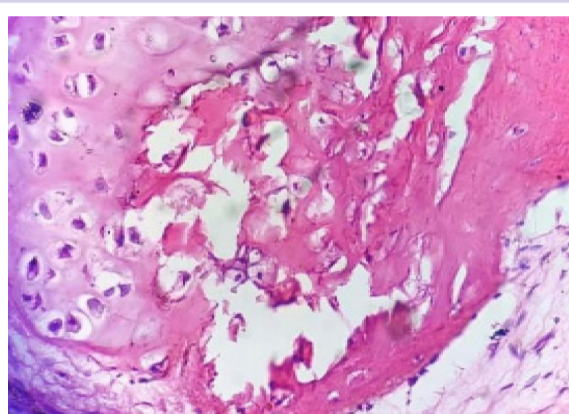


Figure 4: Biopsy revealed epithelial squamous metaplasia, degenerating lymphocytes, histiocytes and few neutrophils in the subepithelial stroma; Osteocartilagenous fragments in the submucosa which were suggestive of Tracheobronchopathia osteochondroplastica

associated with sputum that was rusty coloured, mucopurulent and non foul smelling and was not associated with haemoptysis. Shortness of breath was gradual in onset, grade II according to MMRC and was not associated with wheeze, orthopnea and paroxysmal nocturnal dyspnoea. There was no history of chest pain, change in voice, loss of appetite or loss of weight. He had no similar complaints in the past. He was admitted with acute abdomen, diagnosed as intestinal obstruction 3 years back and was treated in a private hospital. There was no significant family history.

General physical examination revealed pallor and a surgical scar was present from below the umbilicus to the pubic symphysis. At the time of presentation his temperature was 99, Pulse Rate: 96/min, SpO₂: 96% at room air, Respiratory rate: 25 cycles/min, BP-110/70 mm/Hg. Respiratory System examination revealed a dull percussion note and coarse crepts in the right infra

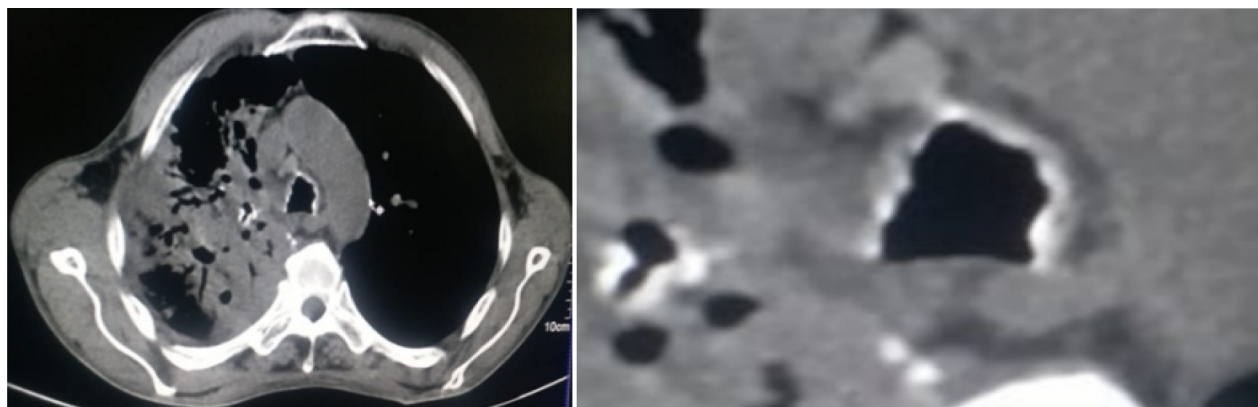


Figure 5: CT Chest showed consolidation with cavities in the right upper lobe, irregular thickening and nodularity of tracheal cartilage with calcification in the trachea and both main bronchi suggestive of Tracheobronchopathia osteochondroplastica



Figure 6: Chest radiograph (posteroanterior view) on follow up after one month showed resolution

clavicular area. B/ L rhonchi were heard in all the areas. Rest of the physical examination was within normal limits. A provisional diagnosis of right upper lobe pneumonia was made and was investigated further.

The blood investigations revealed Hemoglobin of 9.1gm%; TLC23,600cells/cumm; DLC: Neutrophils-74%, Eosinophils- 02%; other hematological and biochemical investigations were within normal limits. Chest radiograph (posteroanterior view: Figure 1) showed a large non homogenous opacity with cavities on the right upper zone suggestive of necrotizing consolidation. Sputum for AFB, CBNAAT were negative. Sputum for Grams stain showed pus cells with gram positive cocci and gram negative bacilli. A diagnosis of Community Acquired Pneumonia was made and he was started on empirical antibiotics.

The patient continued to have symptoms and repeat chest

radiograph (posteroanterior view: Figure 2) after 5 days showed worsening. Bronchoscopy was performed which revealed multiple nodules protruding into the lumen of trachea from the anterolateral wall (Figure 3) and purulent secretions were seen from the right upper lobe. Due to the stiffness of the nodules, a forceps biopsy was difficult to perform. Washings, brushings and biopsy were taken. BAL for AFB, CBNAAT were negative; BAL for Grams stain showed pus cells with gram positive cocci and gram negative bacilli; Culture showed E.coli which was sensitive to Cefotaxime, Amikacin and Imipenem.

Brushings for cytology were negative. Biopsy (Figure 4) revealed epithelial squamous metaplasia, degenerating lymphocytes, histiocytes and few neutrophils in the subepithelial stroma; Osteocartilagenous fragments in the submucosa which were suggestive of Tracheobronchopathia osteochondroplastica. CT Chest (Figure 5) was done which showed Consolidation with cavities in the right upper lobe, irregular thickening and nodularity of tracheal cartilage with calcification in the trachea and both main bronchi suggestive of Tracheobronchopathia osteochondroplastica.

Based on bronchoscopy, radiology and clinicopathology a final diagnosis of Tracheobronchopathia osteochondroplastica (TPO) with post obstructive pneumonia was made. The patient was started on bronchodilators, antiinflammatories and antibiotics based on sensitivity pattern. On follow up, one month later the patient improved clinically with resolution on chest radiographs (Figure 6).

DISCUSSION

Tracheobronchopathia osteochondroplastica was first described in detail by Rokitsansky and Wilms in 1857 after an autopsy. In 1896, Von Schroetter documented the

diagnosis in vivo for the first time by using alaryngeal mirror. In 1897, earliest bronchoscopic description of TPO was given by Killian.

The term of 'tracheopathia osteoplastica' was proposed by Aschoff in 1910. It was expanded by Landsberg as 'tracheobronchopathia osteochondroplastica'. With the widespread use of bronchoscopies in the modern era, TPO has become a relatively common disorder. However, because of misdiagnosis, underreporting and ignorance, the incidence of TO is only 0.3% at autopsies and 1/125 to 1/5000 with bronchoscopies.^[1]

TPO is a rare disorder with a benign course. It is characterized by the accumulation of diffuse cartilaginous and osseous nodules protruding into the mucosa of the trachea and bronchus. As the nodules originate in the airway cartilages, the posterior membranous wall of the airways is typically spared. TPO occurs most commonly between fourth to sixth decades of life. There is no sex predominance. Chroneou et al reported that men are more at-risk than women.^[2]

The etiology of TPO remains unknown. Several theories addressed the occurrence and development of TPO. It was estimated that a slowly progressing inflammatory process of the mucosa caused squamous metaplasia of the epithelium, damaged the normal architecture of the airway, affected the defense mechanisms, especially the efficiency of mucociliary clearing respiratory secretions and led to recurrent infections at last.^[3]

Zack and Rozenshtein^[4] noted that undifferentiated connective tissue cells situated in the submucosal and elasticity layer transformed to cartilage and bone. Ecchondrosis and exostosis put forth by Virchow in 1869 and metaplasia of the elastic tissue by Aschoff in 1910 are the two main theories adopted in numerous publications.^[5] Recently, bone morphogenetic protein-2 (BMP-2) and transforming growth factor beta-1 (TGF- β 1) were considered the potent inducers for new bone formation.^[6]

TPO can be asymptomatic or associated to non-specific respiratory symptoms, such as chronic cough, dyspnoea, hemoptysis, stridor, and recurrent and slowly resolving pneumonia.^[7,8] A retrospective cohort study of 22 patients with TPO 9 showed that chest X-ray was positive in only 16.6 % of cases, while 81.2 % of patients had abnormalities on chest CT scan. These were characterized by irregular tracheobronchial wall thickening, calcification, and, more rarely, tracheal stenosis. Multiple submucosal nodules, with or without calcifications, sparing the posterior membranous wall, and deformation of the cartilaginous tracheal rings in absence of external compression, are pathognomonic findings at CT scan.^[9]

Our case demonstrates how the TPO can present with clinical symptoms of pneumonia and parenchymal consolidation. In a recent study by Zhu et al^[9] recurrent respiratory infections were observed in 8 out of 22 patients (36.3 %); the same percentages have been previously reported by Nienhuis et al.^[8]

The definitive diagnosis is made by bronchoscopy. The typical bronchoscopic findings were described vividly as a cobblestone, a beaded or stalactitic cave, a rock garden, or a veritable mountainscape appearance.^[10,11] In our case, bronchoscopy was done to isolate the pathogens which were responsible for the infection.

Bronchoscopy revealed multiple nodular lesions with a hard texture in the anterolateral wall and purulent secretions were seen from the right upper lobe. In 2004 Dutau et al.^[12] proposed a disease severity classification based on the extent of endoscopic lesions: Stage A: Scattered Nodules (few nodules with large areas of normal mucosa in between); Stage B: Diffuse Nodules (many nodules affecting the entire mucosa, without areas of normal mucosa); Stage C: Lesions Confluent (fusion of adjacent lesions).

In Stage C confluent lesions can lead to a severe breathing impairment due to mechanical obstruction. In unclear cases, bronchial biopsies allow the differential diagnosis of diseases such as amyloidosis, sarcoidosis, papillomatosis, tracheobronchial calcinosis, endobronchial tuberculosis, post-tuberculosis calcified lesions, granulomatosis with polyangiitis and relapsing polychondritis.

In our case, biopsy revealed epithelial squamous metaplasia; Osteocartilagenous fragments in the submucosa allowing us to make a certain diagnosis of TPO. Spirometric tests can be used to evaluate obstructive pattern in symptomatic patients with extensive disease.^[13]

No specific treatments are recommended. Most patients receive palliative treatments, such as antiinflammatories and antitussives. Therapy includes prompt treatment of pulmonary infections with appropriate antibiotics. When a patient presents with acute breathlessness, inhalation of beclometasonedipropionate and budesonide can quickly ease his or her symptoms.^[14]

Endoscopic and surgery treatment is reserved for cases of major bronchial obstruction and include the resection of tracheal segment, partial laryngectomy, laser removal of nodules, rigid bronchoscope dilation, and stent placement (T-Y tube).^[12] Bronchoscopy laser vaporization is the most exact curative treatment.^[15]

CONCLUSION

TPO is a rare disease with benign potential. Poor specificity is observed in clinical manifestation and laboratory investigations. However, a CT scan can show central airway mucosal irregularities and calcification. A bronchoscopy and histopathologic examination greatly contribute to a definitive diagnosis. No specific treatments are recommended, except treatments to alleviate symptoms. Sometimes a therapeutic bronchoscopy or tracheotomy is used to treat severe dyspnea, thereby improving the quality of life.

CONFLICT OF INTEREST:

The authors declared no conflict of interest.

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