CASE REPORT

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A Rare Case of Tracheobronchopathia Osteochondroplastica in Inflammatory Bowel Disease

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Abstract

Tracheobronchopathia osteochondroplastica (TO) is a rare, benign disorder characterised by cartilaginous/osseous nodules in the airways. Its aetiology is not known but various associations have been reported. We report an association of Inflammatory Bowel Disease with TO for the first time. The patient also had organising pneumonia and bronchiectasis with bronchocoele that are known associations with IBD and suggest an autoimmune link between IBD and the pulmonary findings.

Introduction

Tracheobronchopathia osteochondroplastica (TO) is a rare, benign disorder characterised by cartilaginous/osseous nodules in the lower trachea and upper bronchi with occasional involvement of larynx and subglottis, detected in approximately 1 in 2,000 bronchoscopies[1]. TO was historically found incidentally in autopsies but has been increasingly diagnosed with the evolution of flexible bronchoscopy and CT[2]. Its differential diagnoses include infectious deposits, sarcoidosis, chondromas, endobronchial hamartomas, squamous cell papillomas, and rarely malignant conditions like squamous cell carcinoma and adenoid cystic carcinoma. Tracheobronchial nodules are also associated with inflammatory bowel disease (IBD); however, an association of TO with IBD has not been reported[3].We report a case of a young adult male with active IBD exhibiting multiple pulmonary involvements, including TO.

Case report

A 38-year-old male presented with 18 months of progressively worsening rectal bleeding, accompanied by recurrent episodes of high-grade fever and cough. The patient was admitted to the gastroenterology department for evaluation. His familial, personal, occupational and drug history were not contributory. Colonoscopy revealed procto-sigmoiditis. Biopsy indicated colonic mucosal erosion, ulceration, irregular crypt spacing, and cryptitis, with chronic inflammation in the Lamina Propria without any evidence of granulomas or malignant cells, confirming the diagnosis of Ulcerative Colitis.

The patient was referred to pulmonary medicine for evaluation of fever and cough. A detailed history revealed that he had been having cough for around 5 years which had become productive for a year. His chest X-ray showed a patchy opacity in the left mid and lower zone. CT thorax revealed calcified nodules in the trachea (Fig. 1A), a patch of consolidation suggestive of organising pneumonia in left lower lobe (Fig. 1B) along with bronchiectasis and a bronchocoele in the right lower lobe (Fig. 1C).

The patient underwent Flexible Fibreoptic Bronchoscopy (FOB) under conscious sedation. In the trachea, nodules were present anterolaterally, starting below the subglottic space, extending from the trachea into the right main bronchus (RMB) and the left main bronchus (LMB), sparing the posterior wall suggesting a possible cartilaginous origin (Fig. 1D). The size of nodules decreased supero-inferiorly (Fig. 1E-F). Further, multiple nodules were identified anteriorly in the RMB up to 1 cm from the carina, alongside a solitary nodule in the Right Middle Lobe (RML). Similarly, multiple nodules along anterolateral walls were observed in the LMB up to 2.5 cm from the carina. Additionally, purulent secretions were noted in both the Left Upper Lobe (LUL) and Left Lower Lobe (LLL) bronchi. Bronchial washings were taken from the LUL and LLL. Forceps biopsy was attempted from the tracheobronchial nodules but was hindered by the nodules' hardness; only small pieces were obtained. Cryobiopsy of the tracheal nodules also yielded only mucosa in alignment with the possible osteo-cartilagenous nature of the nodules. Transbronchial Lung Biopsy (TBLB) was taken from the LLL in view of the organising pneumonia pattern on HRCT.

Cytological scrutiny of the bronchial wash specimen, showed the presence of respiratory epithelial cells amid a proteinaceous milieu, accompanied by moderate mixed inflammation and absence of any atypical cells. Pyogenic

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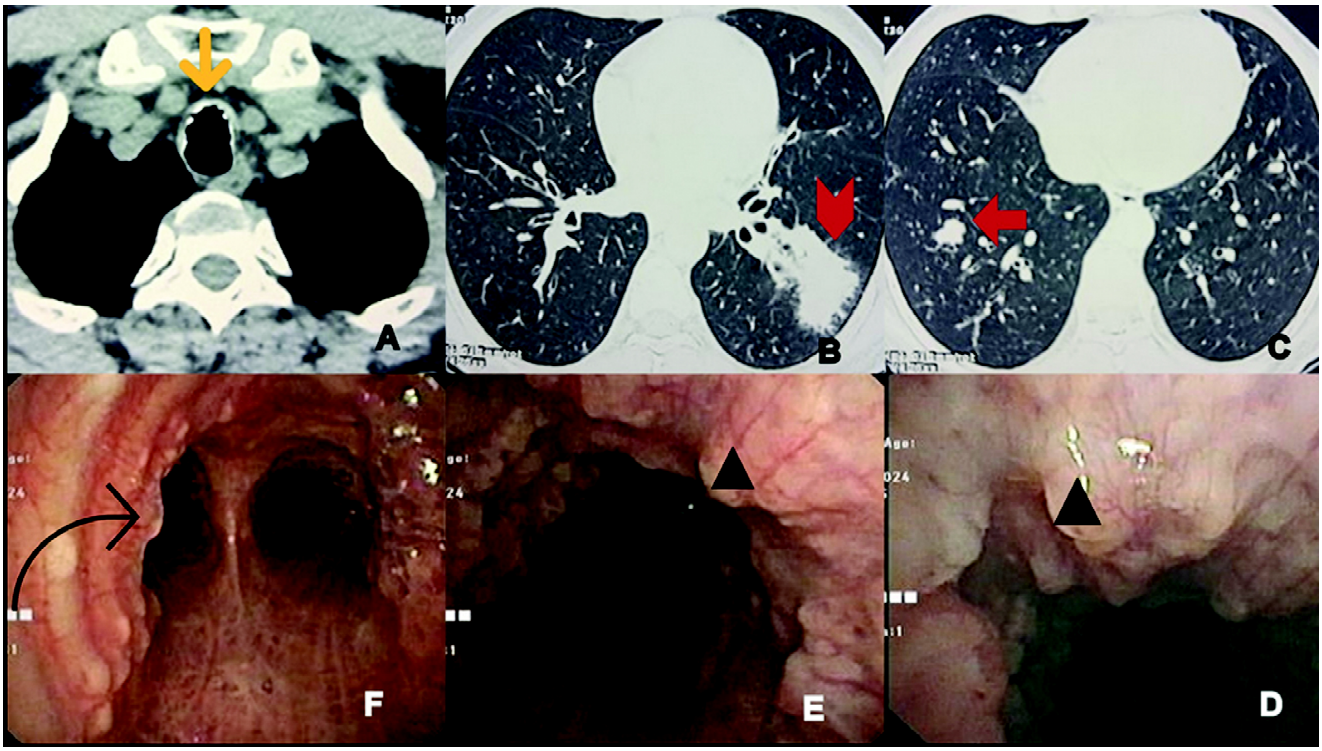

Fig. 1: *Computed Tomography of Chest,* ***A:*** *Mediastinal cut showing multiple calcified tracheal nodules along the anterior surface sparing the posterior surface (Yellow arrow);* ***B:*** *Demonstrates patchy consolidation in the left lower lobe, indicative of organising pneumonia (Red Pointer);* ***C:*** *Shows areas indicating bronchiectasis and Bronchocoele (Red arrow);* ***D:*** *Flexible Fibreoptic Bronchoscopy (FOB) shows large tracheal nodules with cobblestone appearance antero-laterally starting below the subglottic space (Black Triangle);* ***E,F:*** *The nodules were seen to be decreasing in size supero-inferiorly clearly demonstrating their origin along the Tracheal rings (curved Black arrow).*

Table I: Shows the findings from three major case series of Tracheobronchopathia osteochondroplastica.

Author	No of cases	Year	Mean age (years)	M: F Ratio	Location	Chief complaints	Radiological and Spirometry findings	Outcome
Härmä *et al*[5]	30 (over 12 years)	1977	46.5	1:3.6	Finland	long-term recurrent cough, hoarseness, and periodic expectoration	Not studied	Ten cases were incidentally identified via bronchoscopy, 2 through autopsy, and 18 through systemic examination. Among the latter, 10 received preliminary diagnoses via indirect laryngoscopy.
Vivian Leske *et al*[8]	41	2001	63 ± 15	1:1	France	29% had atrophic rhinitis, sinusitis, or pharyngitis and 51% had chronic or recurrent lower respiratory tract disease	74% of Chest CT scans showed submucosal nodulesPFT in 39% of cases was Obstructive, 18% a restrictive defect, and normal spirometry in 43%.	Atypical Posterior tracheal wall involvement occurred in 15% of patients. Initial diagnosis revealed airway stenosis in 10% of patients. Subsequent endoscopic follow-up showed stability in 55%, minimal progression in 28%, and significant progression in 17% of patients.
Ying Zhu *et al*[14]	22	2014	47.45 ± 10.91	1:1	China	chronic cough (n = 14) and increased sputum production (n = 10).	In 18 out of 22 patients, CT revealed findings consistent with TO, including beaded intraluminal calcifications and/or increased luminal thickness Spirometry not studied	Patients were categorised by bronchoscopic severity: Stage I (n = 2), Stage II (n = 6), and Stage III (n = 14). Two patients, treated with inhaled corticosteroids, showed bronchoscopic improvement, indicating potential disease resolution.

culture of the BAL fluid revealed *Pseudomonas* growth. A Ziehl-Neelsen stain for acid-fast Bacilli (AFB) yielded a negative result, Cartridge-Based Nucleic Acid Amplification Test (CBNAAT) for *Mycobacterium tuberculosis* and Potassium Hydroxide (KOH) stain and fungal culture of the BAL fluid were subsequently negative.

Histological examination of the tracheal biopsy specimen showed squamous metaplasia of the respiratory epithelium,

presence of chronic inflammatory cells, fragments of cartilage, and presence of calcified material. No evidence of malignancy was seen. Considering the pattern of tracheobronchial nodules, their consistency, biopsy findings and radiological appearance a diagnosis of Tracheobronchopathiaosteochondroplastica (TO) in association with Ulcerative colitis was made. Transbronchial lung biopsy showed presence of granulation tissue within alveoli with lymphocytic and plasma cell infiltrates with scanty fibrosis, confirming the presence of organising pneumonia. The patient was given antipseudomonal antibiotics as per the culture sensitivity reports of Broncho-alveolar lavage fluid and became afebrile in a week.

Discussion

In 1910, Aschoff[4] coined "Tracheopathiaosteoplastica" suggesting a connective tissue disorder affecting the internal elastic membrane of the trachea and major bronchi. Infective associations with Atrophic Rhinitis, *Mycobacterium avium, Mycobacterium tuberculosis*, and *Klebsiella* are reported[5]. Occupational silica exposure, tissue degeneration, calcium and phosphorous metabolic disturbances, congenital anomaly, chemical or mechanical irritation, primary amyloidosis, familial occurrence, and cold climate conditions in regions like Finland and Northern Sweden are linked to TO[6]. Table I shows the findings of 3 major case series of TO. The harsh climate or irritation contributing to chronic infections, and thereby leading to increased sensitivity of the airway epithelium has been postulated as a possible pathogenetic mechanism[5]. Regardless of the trigger, histopathologic evidence suggests a significant role of bone morphogenetic protein-2, in synergy with transforming growth factor-β1, in nodule formation in the tracheal submucosa[7].

Chronic cough, attributed to mucociliary escalator dysfunction, often lasting 4 - 6 years before diagnosis, is the primary presentation of TO. Other symptoms may include acute pneumonia, hemoptysis, dysphonia, and breathing difficulties[8].

CT is the recommended imaging for TO diagnosis and monitoring, while bronchoscopy is the gold standard, revealing characteristic appearances like cobblestone or stalactitic cave formations[6,9] (Fig. 1A). Forceps biopsy may be challenging due to the nodules' hardness, but histopathologic examination is crucial for confirmation in atypical cases. Cryobiopsy is not suitable as the nodules are osteocartilagenous in nature[10].

The most frequent histopathologic findings are the characteristic presence of bone in bronchial submucosa and squamous metaplasia of the tracheal epithelium.

Our patient had a co-existing biopsy proven IBD. IBD can have pulmonary complications, including bronchiectasis, bronchiolitis, organising pneumonia, necrobiotic nodules, interstitial lung disease and treatment-related complications[3]. Tracheal nodules, in TO are usually calcified nodules in the cartilaginous trachea sparing posterior membrane, while in IBD, there is diffuse or focal tracheal narrowing[11]. This is the first reported case of concurrent TO and IBD, suggesting a possible autoimmune link. Routine bronchoscopy for cough in IBD patients and IBD evaluation for incidentally found TOs during bronchoscopy may elucidate their association. IBD rarely involves the lungs with various pathologies, implying a potential connection between TO and IBD-related pulmonary manifestations such as organising pneumonia and bronchocoele[12].

Treatment for TO ranges from providing symptomatic relief to more invasive bronchoscopic interventions for severe airway obstruction. While inhaled corticosteroids, such as Budesonide, have shown efficacy in managing inflammation associated with TO, their effectiveness is limited when it comes to addressing osseous lesions. Further research is imperative to establish optimal dosage and treatment duration, especially in cases involving bony lesions. Options such as laser ablation, mechanical debulking, and surgical resection of the affected airway exist, but there remains a lack of specific guidelines for the management of this condition[13].

Conclusion

IBD is known to have various pulmonary complications including tracheobronchial nodules. We report a case of TO occurring in IBD for the first time. In patients of IBD presenting with chronic cough, TO should be suspected. Also, patients with TO should be evaluated for IBD. This report highlights a potential association between TO and IBD and mandates further studies.

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