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Tracheobronchopathia Osteochondroplastica (TPO) - An Unusual Presentation

Agarwal SV*, Gupta ML and Yadav SS

Department of Pulmonary Medicine, Santokba Durlabhji Memorial Hospital Cum Medical Research Institute, India

***Corresponding author:** S Vinayak Agarwal, Department of Pulmonary Medicine, Santokba Durlabhji Memorial Hospital Cum Medical Research Institute, Jaipur, Rajasthan, India, Tel: 0141 236 5415; E-mail: svinayak.agarwal8@gmail.com

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Abstract

Tracheobronchopathia osteochondroplastica (TPO) is a rare and usually benign disorder affecting the trachea and occasionally the bronchus. We describe the case of a 50-year-old female who was discovered to have TPO. This case was also unusual since the patient presented with acute hypercapnic respiratory failure and had no previous symptoms despite presence of extensive endotracheal lesions.

Keywords: ***Tracheobronchopathia osteochondroplastica (TPO); Rare disease; Hypercarbia; Fibreoptic bronchoscopy (FOB)***

1. Introduction

TPO is an idiopathic non-malignant disease involving large airways having characteristic features of focal or diffuse sub mucosal cartilaginous to osseous nodules overlying the cartilaginous rings projecting into the lumen of larynx, trachea or bronchi, with sparing of the posterior membranous trachea [1]. The clinical presentation varies from an incidental to central airway obstruction with chronic cough, dyspnea on exertion, and hemoptysis being the most common symptoms [2]. Even though radiological imaging studies may indicate the diagnosis, bronchoscopy is the most definitive diagnostic test. Recognition of its features on bronchoscopy is thus important for all involved in its management and diagnosis. Rare presentation include fever, dryness of throat, weakness, hoarseness, wheezing, stridor. Our case illustrates another rare manifestation in which the patient of TPO presented with acute hypercapnic respiratory failure.

2. Case Report

A 50-year-old, non-smoker female presented to the emergency department with the chief complaints of dyspnea and cough with duration of 6 days. At presentation patient was having low oxygen saturation (spo2) of 85% on oxygen mask with blood

pressure of 148/72 mmHg and pulse rate of 106/min. Arterial blood gas analysis (ABG) revealed acute hypercapnic respiratory failure with pH 7.35, pCO2 91.2 mmHg, pO2 48.1 mmHg hence she was intubated due to increasing respiratory distress and increased retention of carbon dioxide. No wheezing was noted on auscultation.

No history of any respiratory related symptoms was present in the patient prior to the onset of dyspnea 6 days ago. No neurological or cardiovascular related history was present. History of allergies was unknown and there were no significant abnormalities on physical examination. Patient denied history of fever, night sweats, weight loss, or hemoptysis. Patient had known comorbidities like hypertension and type 2 diabetes mellitus. Computed tomography (CT) chest of the patient showed bilateral mild pleural effusion along with calcified nodules seen on anterior and lateral wall of trachea (FIG. 1). Further fibreoptic bronchoscopy (FOB) was performed which revealed multiple nodules of varying size present over antero-lateral wall of trachea, with relative sparing of the posterior membranous wall (FIG 2).

Tracheal secretion was negative for bacteria, fungi and mycobacteria. Cultures were negative for bacteria. BAL did not reveal any atypical cells. Due to high pressure support it was difficult to extubate. Hence patient was tracheostomized and was later decannulated and discharged.

Acute hypercapnic respiratory failure in our patient could have been due to:

1. narrowed airways
2. retained mucous secretions in tracheobronchial tree
3. Could have been due to obstructive airway disease as patient was of rural background with history of chulha exposure

Spirometry could not be performed in the patient as she on mechanical ventilation initially and was later tracheostomized.

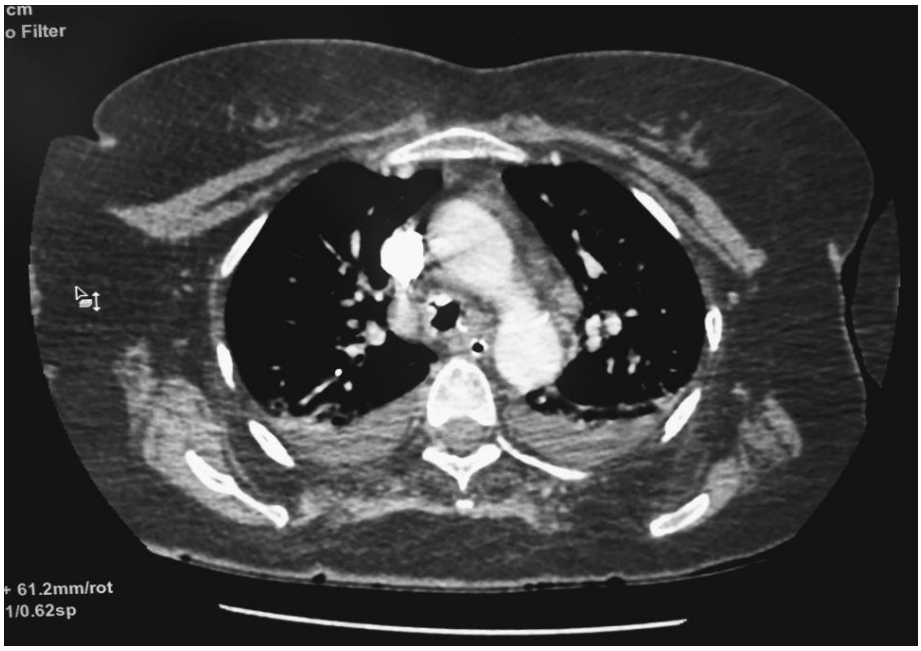

FIG. 1. **CT Chest showing bilateral mild pleural effusion along with calcified nodules seen on anterior and lateral wall of trachea.**

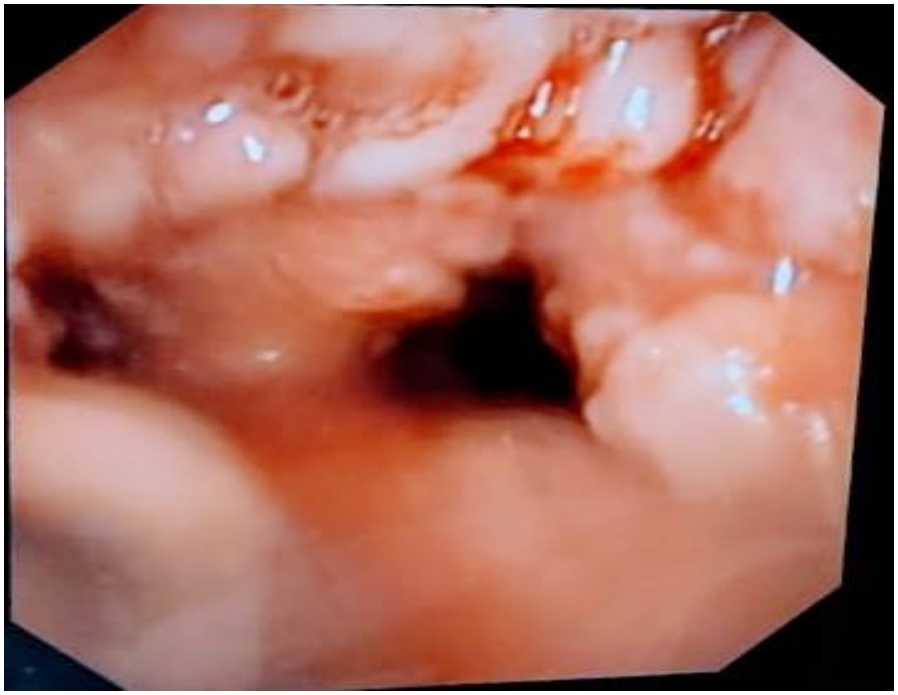

FIG. 2. **FOB revealed multiple nodules of varying size present over antero-lateral wall of trachea, with relative sparing of the posterior membranous wall.**

3. Discussion

TPO first described in 1857 by Wilks but it was in 1897 that the earliest bronchoscopic description of TPO was given by Killian. Till date up to more than 500 cases of TPO has been reported worldwide but the true incidence of the disease is considered to be higher due to its absent or nonspecific symptoms. The detection rate of TPO ranged from 1:400 (0.25%) to 3:1000 (0.30%) in autopsy studies and 1:125(0.80%) to 1:1000(0.01%) via bronchoscopy [2-4].

The etiology of TPO still remains unknown but their associations with inflammation, trauma, chronic infections, amyloidosis and silicosis have been hypothesized. Cases have been reported in association with allergic rhinitis. Prakash et al. [5] reported a familial occurrence of TPO. A Study by Tajima et al. [6] showed that bone morphogenetic protein-2(BMP-2) and transforming growth factor beta-1(TGF-beta 1) are potent inducers of new bone formation.

Usually, it presents between the fourth and the seventh decades of life [2]. Although no gender predominance has been described but a slight male predominance has been noted by many investigators [2,4].

Most patients with TPO are asymptomatic. Symptoms usually correlate with the degree and site of airway obstruction. Symptoms are nonspecific and may include chronic cough, dyspnea, hemoptysis, wheezing and recurrent respiratory infections [4]. Due to its slow progression and the non-specificity of the symptoms, it can be misinterpreted as allergy or asthma. The lack of wheezing or history of atopy and the rapid resolution of hypercarbia with endotracheal intubation were suggestive of upper airway disease.

In our patient dyspnea and cough was the presenting feature along with acute hypercapnic respiratory failure. Although dyspnea and cough have been reported as the presenting feature in many cases of TPO but there has been only one such case of acute hypercapnic respiratory failure reported previously.

Chest X-ray is not useful for detecting the nodules and is usually normal in TPO patients. A recent retrospective cohort study of 22 patient conducted by Zhu et al. [7] showed that the chest x-ray was positive in only 16.6% of cases, while 81.2% of patients had abnormalities on Chest CT scan. CT of the chest reveals characteristic multiple calcified projecting nodules, with or without tracheal wall thickening and lumen stenosis. The unique geographical distribution of TPO nodules, typically sparing the posterior tracheal wall, is useful in distinguishing TPO from other nodular conditions affecting the large airways, such as tuberculosis, neoplastic diseases, endobronchial sarcoid, and amyloidosis.

Bronchoscopy and pathological examination are still the gold standard for diagnosing TPO. The typical bronchoscopic findings of TPO are multiple isolated or converging nodules measured between 1 mm and 10 mm protruding into the airway lumen. These lesions are commonly found in the distal 2/3 of trachea. Proximal trachea, subglottic region and larynx involvement have also been demonstrated [8].

In our patient nodules were located throughout the mucosa of trachea, sparing the membranous trachea which helped in reaching to the diagnosis of TPO.

There is no specific treatment for TPO. Intubation may be difficult because of the calcified rings of trachea. Occasionally tracheostomy is required. The treatment varies from symptomatic management, bronchoscopic intervention to operative correction depending on the severity of the airway obstruction. Resection of the affected tracheal segment, anterior laryngo fissure, partial laryngectomy, laser removal of the nodules, rigid bronchoscopic dilation and stent placement are reported surgical options.

4. Conclusion

TPO is a rare disease having a benign course with the potential to provoke acute respiratory failure. This case report highlights that acute hypercapnic respiratory failure can be a presenting feature in patients with TPO hence should be considered as one of the differential diagnosis during evaluation.

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