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Tracheobronchopathia Osteochondroplastica - A Rare Association with Malignancy

KEYWORDS: Tracheobronchopathia osteochondroplastica (TBO), Bronchogenic carcinoma.

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ABSTRACT *Tracheobronchopathia Osteochondroplastica (TBO) is a rare benign disorder characterised by multiple submucosal nodules of trachea and bronchi. Most cases are asymptomatic but few cases are described in association with lung malignancy. So far only one case has been reported from India in association with lung malignancy. This is a case report of TBO detected as an incidental finding during bronchoscopy, while investigating a mass lesion of lung.*

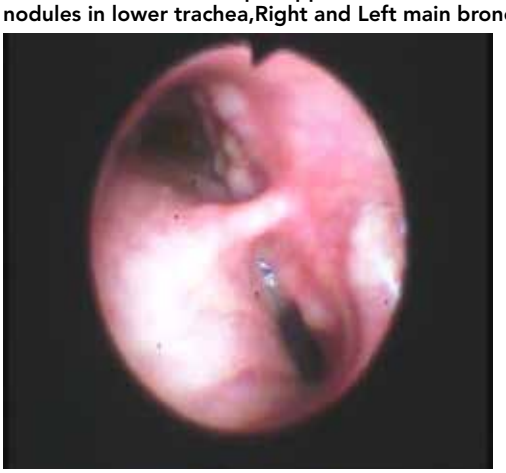
INTRODUCTION:
TBO is a rare condition of major airways where multiple cartilaginous, osseous submucosal nodules are projected into lumen. Although It's considered to be benign & asymptomatic, it can lead to refractory cough, noisy breathing, haemoptysis, difficult intubation, respiratory insufficiency. No definite relation has been established with malignancy but there are several case reports in association with this entity (3) .Here we present a case of TBO which was detected during evaluation of a lung mass lesion.

CASE REPORT:
An 80 year male patient presented with cough, severe left chest pain of 2 months duration, breathlessness of 10 years duration. Breathlessness was of grade III severity, progressive in nature. His chest pain was on left posterior aspect, dull in nature and nonradiating. He had diabetes mellitus, hypertensive on regular medication. He was using on & off bronchodilators for breathlessness. He was a chronic smoker of 50 pack years. Clinical evaluation showed diminished breath sounds on interscapular area and expiratory rhonchi on both sides .Biochemical evaluation showed mild anemia .Chest X ray showed a mass lesion on left upper zone and infiltrates in left lower zone.CECT thorax confirmed mass lesion with calcifications in the left upper lobe, left lower lobe infiltrates.

FIGURE 1: CT scan of thorax showing Left upper lobe mass lesion with calcification.

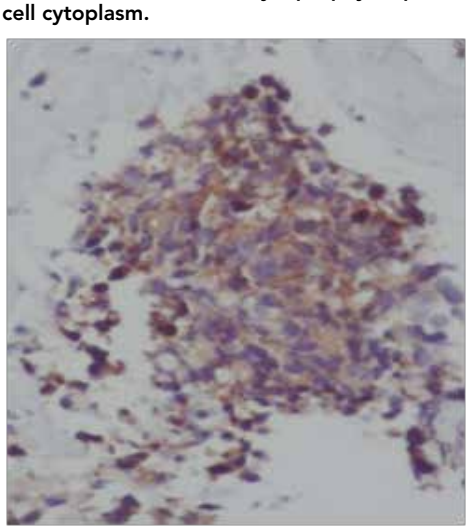
Fibreoptic bronchoscopy was performed which showed multiple nodules with intact mucosa along the anterolateral walls of lower trachea and bronchi.

FIGURE 2: Bronchoscopic appearance of TBO showing nodules in lower trachea,Right and Left main bronchi.



Biopsy was taken from nodules.HPE and cultures no evidence of granulomas, amyloid, malignancy or infections. Histopathological examination of ultrasound guided biopsy of the lung mass revealed small cell carcinoma.

FIGURE 3: Oval to spindle shape cells with scant cytoplasm and hyperchromatic nuclei(40x) (Papanicolau & Giemsa stain) .IHC with Synaptophysin positive in tumor cell cytoplasm.



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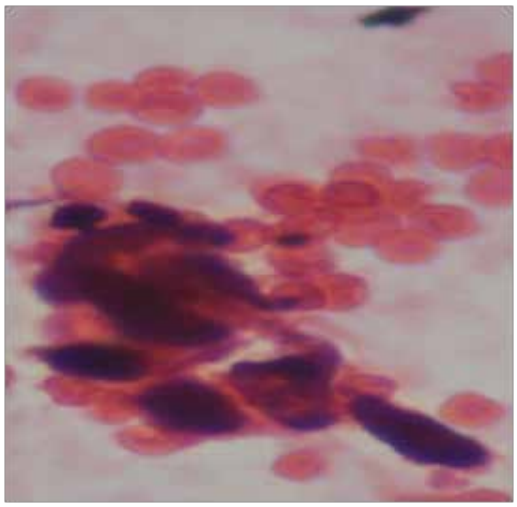

A diagnosis of bronchogenic carcinoma of small cell carcinoma variant, with TBO was made and patient was started on treatment

accordingly.

DISCUSSION:

TBO is a rare benign disorder characterized by osseous and cartligenous nodules in trachea and bronchi covered by normal mucosa (11).First case report was done in 1857 by Wilks during autopsy of a man who died of tuberculosis who described it as "ossific deposits of larynx, trachea, bronchi"(31). Although its true incidence is unknown autopsy series have shown it to be around 1 in 400.As an incidental finding in bronchoscopy, overall incidence reported is around 0.11 %(27) .TBO is a disease of elderly male but it has been reported in young patients also. Median age of presentation is 51yrs (range 16 -68yrs) (2).Many patients are asymptomatic and other clinical presentations include cough(54%)(1,9,15),noisy (10,30), haemoptysis(7,17,19), obstructive pneumonia(1), middle lobe atelectasis(4), recurrent chest infections (21,6), dysphonia(1), tracheal stenosis(1,35), difficult intubation(22,23), progressive respiratory failure (36). Other rare associations with this entity are botryomycosis(8) ,thymoma (12),ruptured hydatid cyst into bronchi,(37)IgA deficiency (18), Ankylosing spondylitis(24).Role of infections like Klebsiella ozanae in association with atrophic rhinitis, atypical mycobacteriae (1), mycobacterium tuberculosis (1,18)have been proposed but not proven. Associated tumours reported were squamous cell carcinoma (25), carcinoid tumour endobronchial tumours, extrapulmonary tumours (27).In our case it is small cell carcinoma .To our best of knowledge no case of small cell carcinoma has been reported in association with TBO in India. Pathology includes multiple osseous, catilagenous nodules in the submucosa of major airways with intact mucosa .These osseous tissues may show marrow suggesting that these can be areas of active haematopoiesis.These ossifications may connect to the perichondrium of cartilage(1).One case report has described lesions as "Soft TPO",where lesions were red and soft (16) .Amyloid tissue has been reported in some cases(1,5). Nodules are present in the anterolateral walls of airways may occlude the airway lumen but classically there is sparing of posterior membrane .Various hypotheses have been proposed regarding pathogenesis,include metaplasia of elastic tissue(1), exostosis and ecchondrosis of the cartilage of tracheal rings .Role of Bone Morphogenic Protein -2 and TGF beta -1 is described recently(20).These factors act synergistically to induce ossifications. Differential diagnosis include sarcoidosis,amyloidosis,aging related calcifications ,endobronchial metastases and papillomatosis. Amyloidosis classically involves entire entire circumference of airway and doesn't show sparing of posterior wall(32,34). Sarcoidosis is diagnosed by noncaseating granulomas and extrapulmonary involvement. Diagnosis is done by bronchoscopic classical appearance of involvement of lower 2/3 of trachea ,major bronchi and presence of ossifications and cartilagenous tissue in biopsy.Various descriptions have been given to the bronchoscopic appearance like "rock garden appearance,,cauliflower appearance,"(26).Imaging like chest X ray ,CT scan , MRI may suggest but bronchoscopy is necessary for diagnosis . Helical CT scan shows diffuse narrowing of trachea and bronchi due to lobulated overgrowth of submucosal tissues, associated calcifications(1,13,14). Spirometry shows mostly obstructive pattern but may show restrictive pattern also (35). A classification was proposed based on bronchoscopic appearance to indicate severity of disease-scattered ,diffuse and confluent types(29). Autoflouresence method has been suggested for diagnostic purpose(28). Prognosis depends upon progression of disease, associated condition. Leske 's endoscopic followup of lesions showed that progression of lesions in 45% of cases(1) .One case report showed rapid progression of lesions leading to tracheal stenosis(33)while another was followed for 20 years without worsening of the disease(38).Deaths are reported due to sepsis (1), respiratory failure(36). Management of this entity includes observation, Coring of lesions with the tip of rigid bronchoscope, NdYAG laser photoevaporation(LPE), polyflex stenting, surgical debulking, external irradiation, tracheostomy with long term home oxygenation.Inhaled corticosteroids provided relief in some patients.Although these measures are not for curative intention they provide symptomatic relief and better quality of life.(1,29).

CONCLUSION :

TBO is a rare entity of trachea and bronchi .Although it is a benign condition it's association with malignancy and refractory cough ,hemoptysis makes it a condition to be sought in diagnostic dilemmas. Bronchoscopy provides diagnosis.

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