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Original Research Paper

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TRACHEOBRONCHOPATHIA OSTEOCHONDROPLASTICA: A RARE PRESENTATION AS ACUTE EXACERBATION OF CHRONIC OBSTRUCTIVE PULMONARY DISEASE

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ABSTRACT Tracheobronchopathia osteochondroplastica is a rare benign, idiopathic disease characterized by abnormal chondrification and ossification of cartilages of the large airways. We hereby report a case diagnosed as group D chronic obstructive pulmonary disease which on further evaluation with high resolution computed tomography thorax and bronchoscopy was diagnosed as tracheobronchopathia osteochondroplastica .The disease is characterized by abnormal chondrification and ossification of cartilages of the large airways. The most common presenting symptoms are cough, hemoptysis, dyspnea on exertion, wheezing, or constitutional symptoms like fever with chills because of recurrent chest infections.CT chest and bronchoscopy with biopsy are diagnostic of tracheobronchopathia osteochondroplastica .The most common complication of this disease is recurrent chest infections. Most patients can be managed conservatively with antibiotics, inhaled steroids and bronchodilators. The awareness among clinicians about this disease entity can lead to early diagnosis.

KEYWORDS : Tracheobronchopathia osteochondroplastica, obstructive, tomography, bronchoscopy

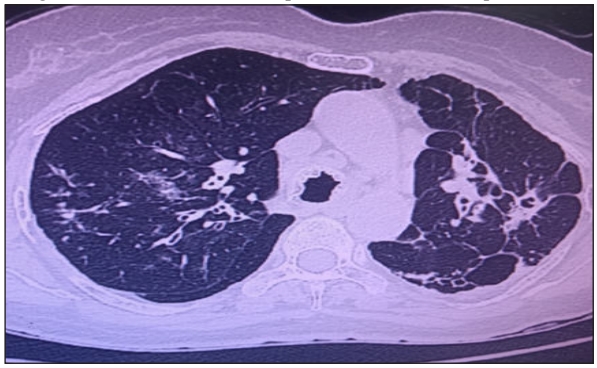
INTRODUCTION

Tracheobronchopathia osteochondroplastica is rare benign disease, first described in 1857 by Wilks and colleagues[1], characterized by abnormal chondrification and ossification of cartilages of tracheobronchial tree with sparing of posterior wall. The most common site of involvement of disease is anterolateral wall of the lower two thirds of the trachea. There is rarely involvement of proximal trachea, subglottic region and larynx. The reported incidence of disease is 0.01 to 4.2 per 100,000 inhabitants[2]. The disease is often diagnosed between fifth to seventh decades of life with no gender predominance[3]. The symptoms of disease depend upon site and degree of airway obstruction. Most patients are asymptomatic or have nonspecific respiratory symptoms. In most patients, disease is stable over years as evident from repeated bronchoscopies. Thereby most cases can be managed conservatively. The most dreaded presentation of the disease is stridor and respiratory failure and these patients have difficult airways. Most cases are misdiagnosed as asthma for years. Thereby awareness among clinicians about this disease is must. The differential diagnoses to be considered when tracheal wall nodules are diffuse and involving the posterior tracheal wall are amyloidosis, sarcoidosis, papillomatosis, tracheobronchial calcinosis and post tuberculosis calcified lesion. The review of literature shows that there is no increase in incidence of malignant disease in patients with tracheobronchopathia osteochondroplastica.

CASE REPORT

A 63 years old lady, non-smoker with previous history of type II diabetes and hypertension presented in emergency department of our medical facility with chief complains of cough breathlessness on exertion, hemoptysis and fever since 4 to 5 days. She had history of similar episodes in past and was diagnosed as a case of group D chronic obstructive pulmonary disease. She was on inhaled corticosteroid, beta2 agonist and anticholinergics for 15 years. The previous pulmonary function test revealed severe fixed obstruction with no bronchodilator reversibility. On examination, she was drowsy but arousable with Pulse - 110 beats/ min; BP - 110/68; Respiratory rate -40/min; SpO2 -89% on room air and temperature -100 F. The patient had audible wheeze. All relevant blood investigations, echocardiography and high resolution computed tomography thorax were done. The arterial blood gas analysis showed compensated type 2respiratory failure. She was managed in medical ICU with broad-spectrum antibiotics, oxygen, non invasive ventilation, bronchodilator and other supportive treatment. The blood investigations revealed neutrophilic leukocytosis with procalcitonin of 4 ng/ml.

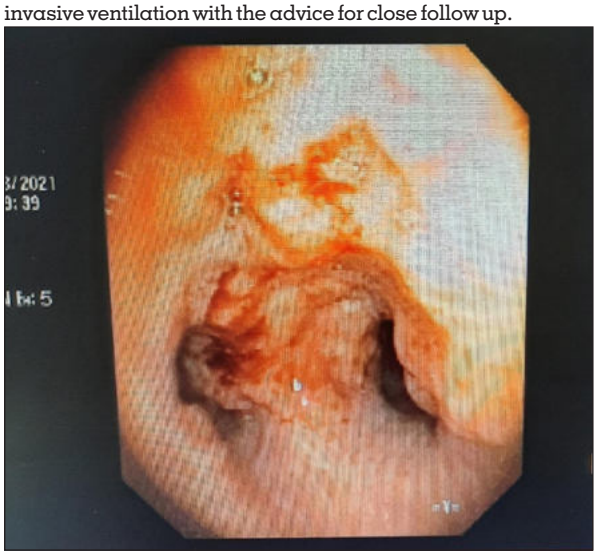
Liver function test, kidney function test and serum IgE level were within normal limits. The electrocardiogram and echocardiography ruled out any cardiac disease. Chest X ray showed bilateral non-homogenous opacity. Her hemoptysis improved with the given treatment .The blood and urine culture of patient was sterile and sputum culture grew Klebsiella pneumonia which was sensitive to cefoperazone sulbactam started empirically. The high resolution computed tomography thorax showed thickening and irregularity of the tracheal wall with calcification suggestive of tracheobronchopathia osteochondroplastica with bronchiectasis and centrilobular nodules in bilateral lower lobes (Picture-1). Bronchoscopy with lavage was planned to rule out tracheobronchopathia osteochondroplastica and tuberculosis. Flexible fiber optic bronchoscopy picture was diagnostic of tracheobronchopathia osteochondroplastica.



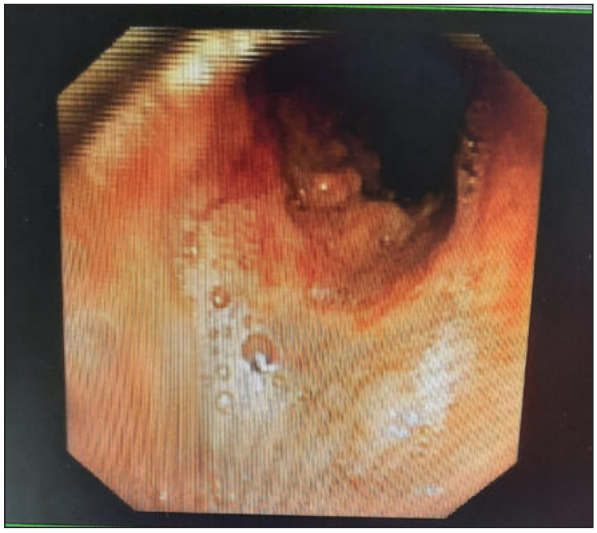
Picture-1: Thickening and irregularity of the tracheal wall with

calcification suggestive of tracheobronchopathia osteochondroplastica. Also there is bronchiectasis and centrilobular nodules in bilateral lower lobes

A grating sensation was felt on passage of bronchoscope in trachea. There were multiple diffuse nodules of 5-10mm in size in distal part of trachea with few showing ulceration with sparing of posterior wall (Picture-2 &3). The tracheobronchial tree lumen was so narrow that bronchoscope could not be passed beyond carina. Biopsy was attempted, but the sample was non-representative and non-contributory for the histopathological diagnosis. Bronchoscopic picture was diagnostic. Thereby this is a rare case of tracheobronchopathia osteochondroplastica presenting as acute exacerbation of obstructive airway disease. She was prognosticated about her narrow airways and its complication. She improved with given treatment and discharged on inhaled bronchodilators and home non invasive ventilation with the advice for close follow up.



Picture-2 : Bronchoscopy View Of The Lesion; Posterior Wall Of Trachea Is Spared



Picture-3: Bronchoscopy View Of Lesion; Pebbles Stone Like Appearance

DISCUSSION

Tracheobronchopathia osteochondroplastica is a rare benign, idiopathic disease characterized by abnormal chondrification and ossification of cartilages of the large airways. Ecchondrosis and exostosis nodules can cause chronic inflammation and mucosal metaplasia, stiffness and airway obstruction[4]. The etiology of Tracheobronchopathia osteochondroplastica is unknown. The proposed causal factors are chronic infections (e.g. mycobacteriosis), Klebsiella ozaenea infection, chemical or mechanical irritation (e.g. silicosis, smoking), metabolic abnormalities (e.g. amyloidosis), genetic predisposition and immunoglobulin A deficiency. Due to familial occurrence there is suggested a possible role of bone morphogenetic protein-2 and transforming growth factor □-1 in tracheobronchopathia osteochondroplastica. In the case presented above no causal factor can be identified. The most common presenting symptoms are cough, hemoptysis, dyspnea on exertion, wheezing or constitutional symptoms like fever with chills because of recurrent chest infections[5]. Hemoptysis as reported in above case is a common symptom due to an acute infection, bronchiectasis or ulceration of a nodule. The tracheobronchopathia osteochondroplastica is often misdiagnosed as respiratory signs and symptoms are nonspecific. The presented case was also misdiagnosed as a case of group D chronic obstructive pulmonary disease. Thereby computed tomography thorax and bronchoscopy should be planned in patient with no identifiable causal factors for obstructive airway disease and unexplained frequent exacerbations and hemoptysis. Chest X-ray and pulmonary function test are nonspecific and not diagnostic of tracheobronchopathia osteochondroplastica as evident from above case findings. The findings in pulmonary function test can range from normal spirometery to obstructive pattern. In case of significant tracheal lumen narrowing there is flattening of inspiratory and/or expiratory flow-volume loop. Computed tomography thorax and bronchoscopy with biopsy are diagnostic of tracheobronchopathia osteochondroplastica[6]. The most common computed tomography (CT) chest findings are multiple sessile submucosal nodules with or without calcifications, with posterior wall of tracheobronchial tree being spared. The tracheal rings are deformed without any external compression. The bronchoscopic view of bony or cartilaginous nodules measuring between 1 and 10 mm, most commonly in the distal 2/3 of trachea, on anterior tracheal wall with sparing of posterior tracheal wall is gold standard for the diagnosis of tracheobronchopathia osteochondroplastica. Laryngoscopy can be diagnostic if larynx and proximal trachea is involved. In tracheobronchopathia osteochondroplastica biopsies are difficult to take through flexible bronchoscope, but are essential to rule out other differential diagnosis. In presented case bronchoscopic picture was diagnostic and biopsy specimen taken was insufficient for histopathological diagnosis. As per Dutau et al[7] proposed disease severity classification bronchoscopic picture of presented case is of stage B [stage A: scattered nodules, stage B: diffuse nodules, stage C: lesions confluent]. The histopathology findings in tracheobronchopathia osteochondroplastica are submucosal cartilage, submucosal ossification, calcification, hematopoietic bone marrow within the ossified areas and epithelial squamous metaplasia. The most common complication of this disease is recurrent chest infections as seen in presented case. Most patients can be managed conservatively like presented case with antibiotics, inhaled steroids and bronchodilators. More severe cases require surgical and endoscopic treatment like tracheostomy, resection of tracheal segment, partial laryngectomy, stent placement, laser removal of nodules, rigid bronchoscope dilation, and stent placement (T-Y tube)[7].

CONCLUSION:

Tracheobronchopathia osteochondroplastica is a rare benign, idiopathic disease. The awareness among clinicians about this disease entity can lead to early diagnosis. It can also present as stridor and respiratory failure. High resolution computed tomography thoraxt and bronchoscopy should be planned in patient with no identifiable causal factors for

obstructive airway disease and unexplained frequent exacerbations.

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