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ROLE OF CT IN DIAGNOSIS OF TRACHEOBRONCHOPATHIA OSTEOCHONDROPLASTICA

Radiology

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ABSTRACT

Tracheobronchopathia osteochondroplastica is an uncommon idiopathic disease of the tracheobronchial system that may lead to narrowing of the airway lumen from cartilaginous and/or osseous submucosal nodules, which may be focal or diffuse. Due to the lack of awareness of this disease, it remains an under-recognized entity.

The disorder is usually asymptomatic and most of the cases have been diagnosed incidentally at autopsy or during bronchoscopy. We are describing the computed tomography and virtual bronchoscopy findings of two recently diagnosed cases at our institute.

The aim is to perform a detailed review of this rare disease and to familiarize radiologists with imaging appearance of this condition, with the goal of increasing clinical suspicion of this uncommon condition.

KEYWORDS:

Tracheobronchopathia osteochondroplastica (TPO), Computed Tomography (CT), virtual bronchoscopy, tracheobronchial calcifications.

Introduction:

Tracheobronchopathia osteochondroplastica (TPO) is a rare, benign condition involving the lumen of lower two thirds of the trachea and upper part of the main bronchi [1].

The disease is characterised by diffuse nodularities, or polyps, consisting of cartilaginous and/or osseous metaplastic tissue involving the anterior and lateral walls of the tracheobronchial tree and sparing the pars membranacea i.e. posterior tracheal membrane. The nodules are 1 to 3 mm in diameter and may cause narrowing and rigidity of the trachea and bronchi if the nodules are larger in diameters, which is rare [2]. The majority of people with TPO remain either asymptomatic or present with nonspecific respiratory symptoms such as dyspnoea, hoarseness, persistent and often productive cough, haemoptysis, and recurrent or slowly resolving pneumonia [3]. Hence, this condition is usually discovered incidentally during bronchoscopy, or at autopsy or during difficult intubation.

It rarely requires any treatment. As the bronchoscopic finding may be confused with airway malignancy, awareness of this condition is important to prevent unnecessary invasive procedures.

Materials and methods:

The CT scans were done on Philips CT Systems "Brilliance" 64 slice CT scan. The results were analysed on a workstation using Aquarius iNtuition Edition Ver.4.4.8.36.843. Virtual Bronchoscopy was performed using the same workstation.

Case report:

Case 1: A 35 year old male patient presented with dyspnea on exertion since 2 months with no h/o chest pain, cough, tuberculosis or tuberculosis contact.

Chest X-ray revealed an ill-defined inhomogenous radio-opacity in right lower zone with air bronchogram.

CT findings were as follows- Moderate right sided loculated hydro-pneumothorax in lower posterior pleural cavity. Area of consolidation showing multiple cavitatory areas and mucous bronchogram sign within in posterobasal & lateral basal segment of right lower lobe with one of the cavities communicating with the loculated hydro-pneumothorax.

Irregular & nodular submucosal calcification involving the trachea (sparing its posterior membrane) & proximal right main bronchus {Fig 1, 2}. CT generated Virtual bronchoscopy was done which showed nodular surface of the trachea (in place of smooth, regular lumen seen in normal patients {Fig3A}), predominantly along anterior and lateral wall, causing mild luminal irregularity {Fig 3B}.

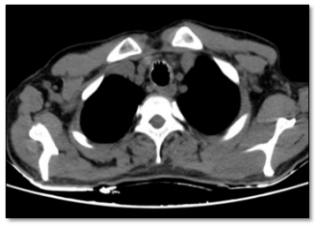

Fig 1 Axial CT Chest image (plain scan) showing anterior tracheal wall irregularity and calcifications.

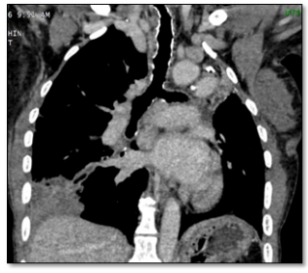

Fig 2 Coronal CT Chest reconstruction (delayed phase) showing wall irregularity and calcifications along lateral wall of distal trachea and right main bronchus.

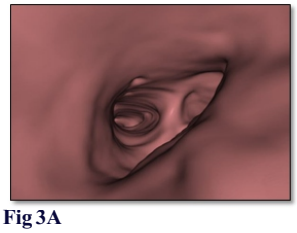

Fig 3A

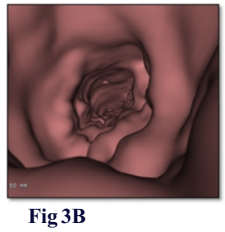

Fig 3B

Virtual bronchoscopic images depicting smooth contour of normal trachea(Fig 3A) and nodular contour in patient 1 with tracheobronchopathia osteochondroplastica(Fig 3B)

Case 2: A 38 year old female patient presented with dyspnoea on exertion grade I which progressed to Grade III-IV within 5-6 days. She

also had history of dry cough and weight loss and past history of tuberculosis.

Chest X-ray showed left upper zone fibrotic changes, inhomogenous radio-opacity with air bronchogram in right lower zone and left middle zone with hemithoracic volume loss.

CT findings were as follows-
A large area of consolidation with air bronchogram was seen in superior and posterobasal segments of right lower lobe. Fibrobronchiectatic collapse of left upper lobe was also seen.

Calcification of tracheal cartilage and major bronchial walls with tracheal contour appearing irregular in appearance {Fig 4, 5}. There was mild wall thickening with irregularity and calcification of anterior wall of left main bronchus and its mild narrowing at origin.

CT generated Virtual bronchoscopy revealed irregular nodular tracheal wall {Fig 6A} extending into left main bronchus {Fig 6B}.

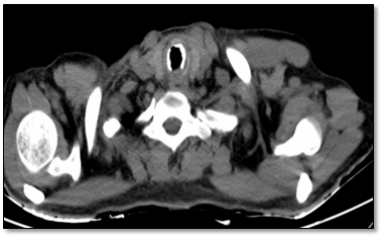

Fig 4 Axial image of plain CT Neck showing anterior wall calcifications and irregularity and nodularity sparing posterior wall.

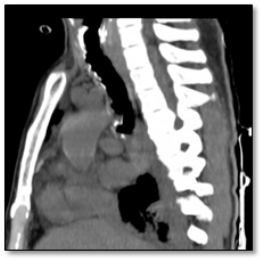

Fig 5 Sagittal CT Chest reconstruction (Plain scan) showing wall irregularity and calcifications along anterior wall of distal trachea

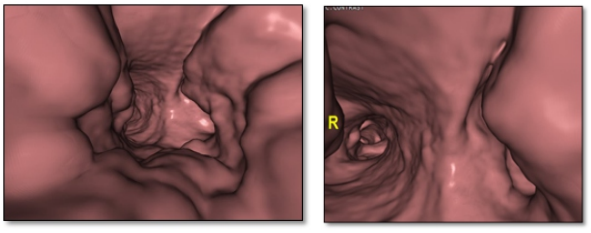

Fig 6A, B Virtual Bronchoscopy of patient 2 showing tracheal as well as bronchial nodularity(smooth, raised beaded nodules)

Discussion:
TPO was first described in detail by Wilks in 1857 [4]. Since then, approximately 300 cases have been reported sporadically throughout the world. The incidence of TPO ranges from 0.01% to 4.2% [5, 6]. There seems to be a higher prevalence in northern Europe, especially in Finland from which about 25% of all cases have been reported [1].

The etiology is unknown. Many presumptive theories have been proposed such as Klebsiella ozaena, genetic factor and inheritance, tissue degenerative process, metabolic disturbance, calcium and phosphorus metabolic disorders, congenital anomaly, chemical or mechanical irritation, primary amyloidosis and malignancy [7]. Amongst these, chronic inflammation is an important one, which was first suggested by Wilks in 1857.

Still the pathogenesis is also unknown; two theories have been suggested regarding the pathogenesis of this disease.

1) First, the Virchow's theory- Ecchondromas are the initial lesions which undergo calcification and ossification leading to nodule formation [8].

2) Second, the Aschoff-Freiburg`s metaplastic theory-The ossification of elastic connective tissue [9].

There is a 3:1 male predilection. Most of the patients are older than 50 years, but cases have also been reported in children [10].

Clinical manifestations of TPO patients are variable and nonspecific. Some patients are asymptomatic; others may suffer from dry cough, productive cough, haemoptysis, dyspnoea, dryness of throat, recurrent lower respiratory tract infection, atelectasis, and difficult intubation.

The PFT (pulmonary function test) of TPO patients are dependent on the severity of TPO. PFT can be within normal range in mild TPO patients or obstructive pattern and flow volume loops in symptomatic patients which may be helpful in the evaluation of follow-up of the course of this disease.

Imaging findings include:
No characteristic appearance of TPO is seen on X-ray. In moderate to severe disease, conventional radiography may reveal tracheal scalloping and nodular irregularity or irregular asymmetric stenosis.

Chest CT scan is the imaging modality of choice for this condition, demonstrates the findings of multiple submucosal calcified nodules involving the anterior and lateral wall of trachea and bronchus, sparing the posterior wall. Although these lesions can be seen anywhere from the larynx to the peripheral bronchi, they more commonly involve distal two third of trachea and proximal bronchi. Since these nodules arise from cartilage, posterior membranous wall of trachea is typically spared (c/w amyloidosis).

CT generated Virtual bronchoscopy is a novel post processing technique that allows non-invasive visualization of the tracheobronchial tree [11]. Although it can show the endoluminal lesions, it cannot assess the mucosal details and tissue samples cannot be obtained. Virtual bronchoscopy is not a replacement for actual bronchoscopy but it can be used as an additional tool during CT evaluation of the tracheobronchial tree. The typical bronchoscopic appearance is of multiple smooth, raised, nodules, often described as "beaded".

Even though imaging studies may give clue to the diagnosis, bronchoscopy and biopsy is the most definitive diagnostic test. The bronchoscopic appearance itself is diagnostic and is characterized by the multiple, varied size smooth whitish nodules, having a 'rock garden' or 'cobblestone' appearance [12]. These nodules are hard on touch and give gritty sensation while passing the scope through the lumen. Biopsy of the nodules is difficult to obtain as forceps slips off of the surface of hard nodules. The characteristic histopathological findings in bronchoscopic biopsies among TPO patients are the cartilage and ossification in the submucosa, calcification, mucosal squamous metaplasia.

Differential diagnosis for calcification of tracheal and bronchial walls are TPO, relapsing polychondritis, amyloidosis, endobronchial sarcoidosis, papillomatosis, tuberculosis, bronchial carcinoma, and tracheobronchial calcinosis and old age.

Amyloidosis unlike TPO, does not spare the posterior wall of the trachea.

Relapsing polychondritis may have a similar distribution as TPO, it characteristically presents with thickened and deformed cartilage with or without calcification, and the inner wall is smooth without discrete intraluminal nodules formation. Also, TPO is not a painful condition, unlike relapsing polychondritis.

Calcification in TPO is much more irregular than the cartilage calcification in healthy old individuals. Biopsy is not essential since the typical appearance of bronchoscopy is sufficient to make a diagnosis. However, histopathological picture may help to rule out other diseases such as mucoepidermoid carcinoma, papillomatosis, sarcoidosis.

The complications associated with TPO include increased risk of

respiratory infections and difficult intubation.
There is no definitive treatment available for TPO, treatment is only offered to symptomatic cases in terms of treatment of infections, bronchodilators, bronchoscopic dilatation(if indicated)[1]. Prognosis is generally good but it is related with degree of airway involvement and luminal narrowing.
Conclusion:

TPO is a rare entity whose true prevalence should be determined. As there are no specific clinical features of the condition with easy detectability on radiological imaging, there is a significant role of the radiologist in identifying the irregular nodular tracheo-bronchial calcifications associated with this condition.

By familiarizing radiologists and pulmonologists with the imaging appearance of this entity, less number of these cases will go unrecognized and will help in prognosticating the complications associated with it.

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