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# Tracheobronchopathia Osteochondroplastica in a 5-year-old Girl

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Tracheobronchopathia osteochondroplastica (TO) is considered an orphan disease with exceptional occurrence in children. We report a 5-year-old female child who was referred to us with chronic cough and recurrent pneumonia. After several investigations, bronchoscopy showed multiple nodules in the tracheobronchial lumen, whose distribution was consistent with TO. The patient was followed for four years, with no change in the pattern of the disease.

**Key words**: *Tracheobronchopathia osteochondroplastica, Recurrent pneumonia.*

Tracheobronchopathia osteochondroplastica (TO), also known as tracheopathia chondro-osteoplastica or tracheopathia osteoplastica, is a rare chronic disease of undetermined etiology. It stems from osteocartilaginous metaplasia and is characterized by an bronchoscopic finding of multiple subepithelial nodules that project into the tracheobronchial lumen and does not affect the lungs or other organs. The nodules may be present from the larynx to the peripheral bronchi in the anterolateral tracheal wall. In general, the posterior wall is spared. This peculiarity is exclusive of this disease and allows the exclusion of other differential diagnoses. TO is often an endoscopic finding and its prevalence in bronchoscopic studies is estimated to range from 1:125 to 1:6000 [1-3]. To date, less than 400 cases have been reported in the literature, and only four were in children [1,2,4].

Diagnosis can be made from the typical endoscopic finding of the disease, dispensing biopsy [1,5,6]. Biopsy, when done, shows ossification or calcification of the bronchial submucosa [6].

## CASE REPORT

A 5-year-old female child with chronic cough and a history of chronic cough and recurrent pneumonia since age 3 years was referred to us. The first episode consisted of right upper lobe pneumonia that progressed to pneumatocele, resulting in a permanent cicatrical atelectasis. The other respiratory infection episodes in the following year were also interpreted as pneumonia but were based, apparently, on a radiographic image that had persisted since the first episode: chronic atelectasis of the left lower lobe and adhesive atelectasis in the right lung. Further examination showed no signs of atopy. Her pulmonary auscultation was normal. The child was HIV seronerative and had a normal immunoglobulin profile. Tuberculin skin test was 6 mm. At the time she was treated as an outpatient and the disease course was favorable, despite frequent coughing and two more episodes of respiratory infection and cervical adenitis treated with antibiotics. At age seven years, she underwent rigid bronchoscopy, which showed widespread nodular mucosal irregularity extending from trachea into bronchial

tree; irregular mucosa, intensely friable when touched by the instrument. The left lower lobe bronchus was not obstructed. Bronchial aspirate was negative for bacteria (including acid-alcohol-fast bacilli and *M. tuberculosis*) and yeasts. The findings were consistent with TO. Biopsy of bronchial mucosa, which did not include cartilage, evidenced a nonspecific, chronic inflammatory process with intense lymphoid hyperplasia in the lamina propria and prevalence of neutrophils. Outpatient care consisted of occasional courses of antibiotics and respiratory physiotherapy. At age 8 years, spirometry was normal. The patient responded well, with occasional, mild respiratory infections. After four years, flexible bronchoscopy showed similar findings (***Fig.* 1**). The patient is now 11 years old, continues to be followed as an outpatient and has a favorable course.

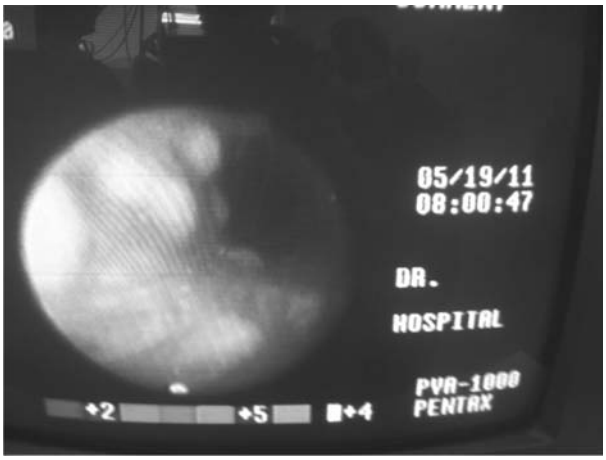


**FIG.1** *Nodular mucosal irregularity of trachea observed on flexible bronchoscopy.*

## DISCUSSION

The present case is relevant because of the rarity of this disease in children. Moreover, TO lesions did not deteriorate after four years of follow-up. The disease may remain stable for a long time or progress insidiously for many years. Rapid progression leading to tracheal obstruction is singular [2,7].

The clinical findings vary. TO may be mistaken for asthma because of the wheezing caused by decreased bronchial diameter. TO may also cause chronic cough [5], dyspnea and hemoptysis [6] but most patients are asymptomatic [1,6].

This patient had chronic atelectasis of the left lower lobe, consequent of recurrent pneumonia. Atelectasis could be explained by obstruction of the bronchial lumen by TO nodules since their promotion of secretion retention established the vicious cycle of atelectasis and pneumonias. The patient did not have asthma, which could be considered an associated condition. However, her wheezing episodes could be attributed to the partial obstruction of the bronchial tree by multiple nodules [2,5,7].

The tracheal and bronchial nodules bled easily and were soft, similar to other reports [1,5]. Since the nodules bled easily during endoscopy, the tissue collected for biopsy during the two endoscopic examinations was more superficial. Hence, the nodules contained no cartilage, only nodular neutrophilic arrangements [5]. This result ruled out other diseases like tuberculosis, hypertrophy of bronchial associated lymphoid tissue, sarcoidosis, etc.

There is no specific treatment for TO. When there is evidence of bronchial infections, the patient should be treated with antibiotics, as presently done. The present patient was never prescribed inhaled corticosteroids, even though they have already been used for TO treatment in some case series [6,7].

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