Tracheobronchopathia asseptica osteochondroplastica in recurrent retrosternal goiter.
Surgical management

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Tracheobronchopathia osteochondroplastica (TPO) is a rare pathology characterized by a progressive segmentary stenosis of the respiratory tract due to proliferation of osteocartilaginous nodules in the lumen of the distal part of the trachea and large bronchial trunks. Prognosis is usually benign, but some cases with an acute progression and a lethal outcome have been described.

Clinical presentation is nonspecific, the chest x-ray is generally normal and there are not typical radiological signs of suspicion: diagnosis of TPO is usually incidental.

We report a case of TPO associated with a retrosternal recurrent goiter. The CT scan conducted to evaluate the extension and the vascular relationships showed the characteristic lesions of the TPO with a segmental stenosis of the trachea greater than 70%. A bronchofiberscopy confirmed the suspect of TPO.

To date, the clinical studies carried out do not show a certain etiology, but all agree that chronic damage or chronic inflammations could be the cause of the onset of structural anomalies of the respiratory tract.

In literature, there is only a report which describes an association between TPO and thyroid pathology. It is obscure whatever these disease could be etiologically or fortuitously associated but a relationship cannot be completely excluded.

Surgeons, anesthetists and radiologists which deal with thyroid pathology must recognize the disease, especially in the presence of bulky retrosternal goiters, to make a correct diagnosis and provide adequate perioperative management.

KEY WORDS: Mediastinal goiter, Osteocartilaginous Nodules, Tracheal Stenosis, Total Thyroidectomy, Tracheobronchopathia Osteochondroplastica

Introduction

Tracheobronchopathia osteochondroplastica (TPO) is a rare pathology characterized by a progressive segmentary stenosis of the respiratory tract with proliferation of multiple submucosal osteocartilaginous nodules in the lumen of the distal part of the trachea and in the large bronchial trunks 1-3.

Diagnosis of TPO is usually incidental, an anaesthesiological check of an unexpected difficulty in tracheal intubation 4 or comes out during computed tomography (CT) or magnetic resonance (MR) chest scan, carried out for different clinical reasons 4-6.

Etiology, pathology and natural history of TPO are unclear. The clinical presentation is nonspecific: respiratory symptoms such as chronic cough, recurrent pulmonary infection, expectorations, and recurrent hemoptysis, sometimes massive, are the most common. Anyway, many subjects do not present symptoms at all. There
are no typical radiological signs of suspicion at chest x-ray, which is generally normal, and there are not morphological and biochemical alterations in the blood tests.

**Case Report**

Male, 69 years old, without a notable past or familiar history. Former smoker since the age of 50, no dusts or chemical occupational exposure. Previous subtotal thyroidectomy for benign goiter at the age of 29. Since the age of 50 progressive evidence of goiter recurrence with predominant extension in the mediastinum rather than in the cervical space, beginning of progressive nocturnal dyspnea. Subsequently the increasing of dyspnea and respiratory discomfort: the patient arrived to our observation.

The CT scan conducted to evaluate the retrosternal extension and the vascular relationships of the goiter showed the characteristic lesions of the TPO with a segmental stenosis of the trachea greater than 70% (Fig. 1).

The bronchofiberoscopy confirmed the suspicion of TPO focusing the typical aspects of osseus and cartilaginous tracheal nodules with normal mucosa (Fig. 2).

**Surgical Treatment**

Total thyroidectomy through a cervical approach. Voluminous retrosternal and retrotracheal right thyroid lobe, while on the left there was a nodular residual tissue. The consistency of the gland was physiological, not ligneous, but with strong adherences between the gland and the tracheal wall. Both laryngeal nerves were identified and saved, two right parathyroids were identified, we didn't found parathyroids in the left residual. The external tracheal wall was intact and without visible or palpable imperfections.

An antibiotic and an anti thrombotic therapy has been done.

**Postoperative Management**

The preoperative patient evaluation allowed to classify the anesthesiologica risk as high (ASA 3), above all for the possibility of respiratory complications. Consequently, a postoperative stay in the intensive care unit (ICU) was planned.

In the ICU, the patient sedation was gradually reduced and an attempt was made to respiratory weaning and extubation after 6 hours.

Immediately after extubation the patient had respiratory failure with ineffective cough, aphonia and desaturation, then it was quickly re-intubated.

The issue of a laryngeal obstruction due to bilateral vocal cord paralysis was ruled out with a videolaryngoscopy performed by removing under sedation the endotracheal tube: a normal motility of the vocal cords and a mild laryngeal edema were observed.

A subsequent bronchoscopy confirmed the characteristics osteochondral lesions with obstruction of the tracheal lumen by 70%, according with the diagnosis of TPO; moreover, it showed that the respiratory failure was...
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Another case-report described the association between TPO and thyroid pathology (goiter or cancer), and this correlation was not confirmed by later reports. In any case, it can not be completely ruled out 14.

caused by purulent pulmonary secretions obstructing the right main bronchus. After a thorough cleaning and removal of secretions for microbiological investigations, antibiotics were implemented, steroid treatment was added, and active humidification of the respiratory system was started.

On the 5th postoperative day a new bronchoscopic procedure was performed to remove secretions, and the patient was extubated. A Ct scan of the chest excluded residual foci of pneumonia.

In the following two days the patient was assisted with noninvasive ventilation until reaching respiratory autonomy, then he was discharged from the ICU and transferred in our surgical unit.

The patient left the hospital in the 16th postoperative day, in good general conditions.

Six months after surgery the patient has normal vocal tones and normal serum calcium, trachea is in axis and has not clinical respiratory discomfort or dyspnea, even if at the CT the nodular tracheal lesions still remain unchanged.

Discussion

TPO is a rare pathology, with no specific clinical presentation, characterized by a chronic worsening course, rarely lethal. Prognosis is usually benign, but some cases with an acute progression and a lethal outcome after weeks or months have been described 7.

The incidence of TPO is uncertain: literature reports a prevalence of 1:400 to 3:1000 cases in autopsy in a various range of population 4,6,7. During bronchofiberoscopy the typical nodular lesions are revealed in 0.017% till to 0.7% of procedures 8,9.

Considering only our cohort of intrathoracic thyroidectomies performed between 2010-2015 our incidence of TPO is 0.6% (1/164).

TPO usually occurs in individuals over 50 years of age, frequently in the sixth or seventh decade of life, with no gender-related differences. There is no evidence of certain risk factors, nor clear relationship with cigarette smoking 1-4,8.

A greater incidence of diagnosis of TPO is reported in the Scandinavian countries, where the atmospheric conditions are severe. Moreover, a greater incidence of diagnosis of TPO is reported in workers with dusts or chemical exposition, suggesting a major pathogenic role of chronic airway damage 10-12.

A recent case-report also suggests a possible role for trauma in the pathogenesis of TPO, documenting an unexpected rapid onset of ossification of the trachea, with lesions similar to those of TPO, in a patient who had an important chest blunt trauma 13.

Another case-report described the association between TPO and thyroid cancer 25. It is obscure whatever TPO could be etiologically or fortuitously associated with a thyroid pathology (goiter or cancer), and this correlation was not confirmed by later reports. In any case, it can not be completely ruled out 14.

Bronchofiberoscopy confirms the suspect of TPO in 90% of cases. The typical aspect of osseus and cartilaginous nodules with normal mucosa is pathognomic. The closest diagnosis is with tracheobronchial amyloidosis in which 22% of patients present tracheopathia usually associated with clinical symptoms suggesting chondrites. The endoscopic aspect of tracheobronchial amyloidosis is different: grayish crumby submucosal plaques and nodules that bleed on contact and extend, differently from TPO, to the posterior wall of trachea 17-19.

Biopsy is not usually performed, because the nodular lesions are very hard and it is not easy to collect material for microscopic examination. However, a biopsy may be needed to exclude other diseases such as neoplasms or papillomas 20.

Actually, there is no specific therapy for this disorder as it is impossible to eliminate all lesions or prevent development of new ones. The treatment of TPO consists in the cure of clinical symptoms and in the prevention of respiratory infections in order to avoid acute illnesses: humidification of the airways, no exposition to irritating agents 21,22, prompt therapy of infection, and administration of bronchodilators ameliorated the clinical condition of some patients. A more aggressive treatment is reported in case of intensive narrowing of the airways to allow an improvement of the lung function tests: the endo lumen nodular lesions can be removed with forcepts through laryngoscopy or bronchoscopy, or with laser ablation and cryotherapy. In case of invalidating stenosis, a stent implantation may be necessary to ensure appropriate breathing. In some patients, improvement is achieved thanks to radiotherapy 23. Surgical treatment in advanced TPO is reported: larynx resection, segmental resection of the trachea, tracheoplasty, removal of changed fragments of the lung 24,25.

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In the case we report the diagnosis was unexpected: the preoperative CT scan performed to study the extension of the mediastinum was not sufficient to diagnose the TPO: there were no evidence of professional dust exposition or previous cervical chest trauma.

Conclusion

We think that surgeons, anesthetists and radiologists which deal with high volume of thyroid pathology need to know the physiopathologic alterations of TPO, and we recommend having a high grade of suspicion for TPO when reporting chest CT/MR.

The pre and post operative treatment for thyroidectomy in patient with TPO need a collective valuation with surgeon, anesthetist, otolaryngologist, pneumologist and bronchoscopist.

In the case we report the diagnostic confirmation of TPO with bronchscopy and the knowledge of its anatomic alterations allowed us to program an effective multi specialized therapeutic treatment.

Riassunto

La tracheobronchopathia osteochondroplastica (TPO) è una patologia rara, priva di una propria presentazione clinica specifica, sovente asintomatica. È caratterizzata dall’insorgenza di una stenosi segmentaria progressiva nel lume distale della trachea e dei bronchi per la genesi di noduli osteocartilaginei sottomucosi. È riportata in letteratura medica una prevalenza tra 1:400 e 3:1000 casi su studi autopsici ed una incidenza tra 0,017% e 0,7% in corso di esami broncoscopici. Non è riportata familiarità. La fascia di età è segnalata differenza di genere, non vi è relazione con il tabagismo, non è riportata professional dust exposure.

La diagnosi della TPO è solitamente incidentale in seguito ad un esame TC/CMR cervico-toracico o, più sovente, è un riscontro anestesiologico in seguito a una intubazione risultata particolarmente difficoltosa. In letteratura medica abbiamo riscontrato un recente case-report che riporta il possibile ruolo traumatico nella patogenesi della TPO, documentando la rapida insorgenza di ossificazione nella trachea con lesioni nodulari similmente alla TPO dopo un importante trauma toracico contusivo. Un solo case report pone invece una possibile relazione tra la TPO e la patologia tiroidea (k papillifero).

Noi riportiamo il caso di TPO associato a un voluminoso gozzo recidivo compressivo retro sternale. L’esame TC effettuato per valutare l’estensione ed i rapporti vascolari dello struma ha evidenziato le lesioni nodulari della TPO con la stenosi segmentaria della trachea del 70%. L’esame broncoscopico ha confermato il sospetto della TPO con il riscontro delle lesioni nodulari ossee e cartilaginee della trachea a mucosa integra.

Il trattamento pre e post operatorio del paziente con TPO è stato sottoposto a tiroideectomia necessitata di una valutazione collegiale del chirurgo dell’anestesista, dell’otorinolaringoiatra del pneumologo e del broncoscopista.

Nel caso report che proponiamo la conferma diagnostica della TPO con la broncoscopia ha permesso di programmare un risolutivo ed efficace trattamento multi-specialistico.

References


