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Surgical management

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Tracheobronchopathia osteochondroplastica in recurrent retrosternal goiter. Surgical management

Tracheobronchopathia osteochondroplastica (TPO) is a rare pathology characterized by a *progressive segmentary stenosis of the respiratory tract due to proliferation of osteocartilagineous 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 non specific, 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 bronchofiberoscopy 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, Osteocartilagineous Nodules, Tracheal Stenosis, Total Thyroidectomy, Tracheobronchopathia Osteochondroplastica

Introduction

Tracheobronchopathiaosteochondroplastica (TPO) is a rare pathology characterized by a progressive segmentary stenosis of the respiratory tract with proliferation of multiple submucosal osteocartilagineous 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 anaesthesiologic 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

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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).

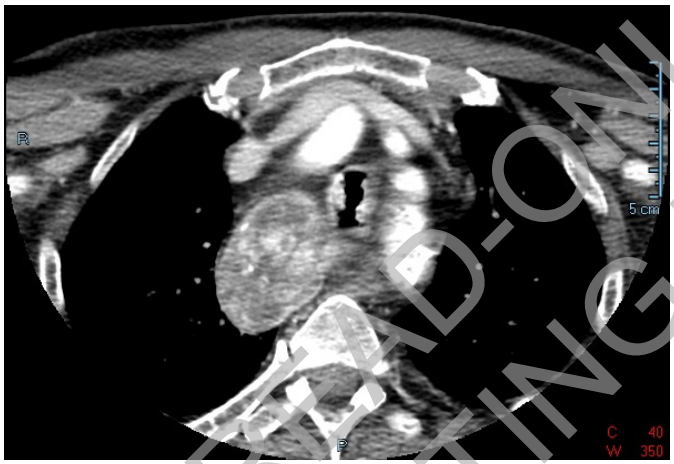

Fig. 1: *CT Image*: recurrent mediastinal goiter in a patient with tracheobronchopathiaosteocondroplastica: proliferation of multiple submucosal osteocartilagineous nodules in the lumen of the distal part of the trachea.

The bronchofiberoscopyconfirmed 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 anesthetesiological 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

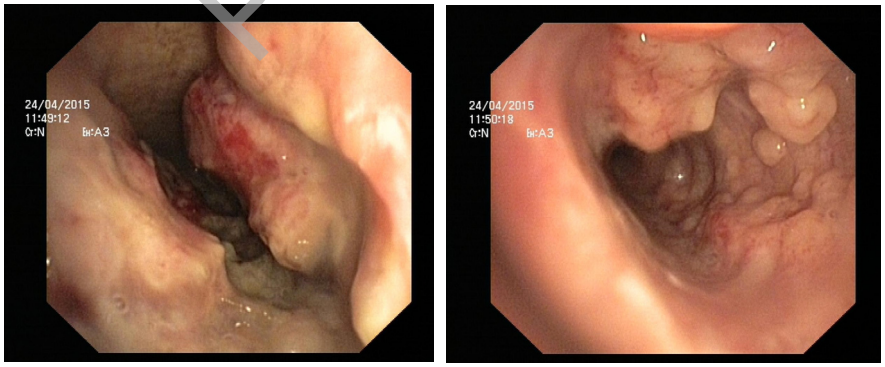

Fig. 2: *Bronchoscopic images*: typical aspects of osseus and cartilaginous tracheal nodules with normal mucosa, pathognonomic for tracheopathiaosteocondroblatica.

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 intrathocacic 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].
The chest x-ray is generally normal and there are not typical radiological signs of suspicion for TPO.
The chest x-ray is generally normal and there are not typical radiological signs of suspicion for TPO. The CT scan is more reliable in the diagnosis of TPO; it usually shows multiple calcified nodules not enhanced after contrast projecting into the lumen and causing segmentary stenosis, with thickening and deformation of the trachea and bronchial wall. Lesions may appear anywhere in the airways, from the larynx down to the peripheral bronchi, but most frequently they occur in the lower 2/3 of the trachea and proximal bronchi. In TPO the posterior part of the trachea is usually free from pathological lesions and this is an important characteristic that distinguishes TPO from other disorders affecting the airways, such as amyloidosis [3,13,15,16].
Bronchofiberoscopy confirms the suspect of TPO in 90% of cases. The typical aspect of osseus and cartilaginous nodules with normal mucosa is pathognonomic. 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 crumbly 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 forceps 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].
TPO shows very little evolution over a period of years and in Leske's[4] series there were no directly imputable deaths. In most cases prognosis is benign, although cas-

es lasting a few months or even weeks with progression of the disease have been reported.
TPO shows very little evolution over a period of years and in Leske's series there were no directly imputable deaths. In most cases prognosis is benign, although cases lasting a few months or even weeks with progression of the disease have been reported [4].
In the case we report the diagnosis was unexpected: the preoperatory CT scan performed to study the extension of the mediastinal goiter was diriment to diagnose the TPO: there were not preoperatory clinical criteria of suspicion, 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 end 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 tracheobronchopathiaosteochondroplastica (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 autoptici ed una incidenza tra 0,017% e 0,7% in corso di esami broncoscopici. Non è segnalata differenza di genere, non vi è relazione con il tabagismo, non è riportata familiarità. La fascia di età colpita è in media superiore ai 50 aa, solitamente in pazienti nella sesta o settima decade di vita.
La diagnosi della TPO è solitamente incidentale in seguito ad un esame TC/RM 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 della 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 broncoscopio 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 da sottoporre a tiroidectomia necessita di una valutazione collegiale del chirurgo dell'anestesista, dell'otorinolaringoiatra del pneumologo e del broncoscopista.
Nel case report che proponiamo la conferma diagnostica della TPO con la broncoscopia ha permesso di programmare un risolutivo ed efficace trattamento multispecialistico.

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