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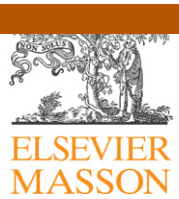

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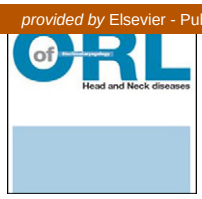

IMAGES

An unusual cause of tracheal stenosis: Diagnosis and management?

A. Bachy[a], N. Saroul[a,*], C. Darcha[b], R. Bellini[c], T. Mom[a], L. Gilain[a]

[a] *Service d'ORL et chirurgie cervicofaciale, hôpital Gabriel-Montpied, CHU de Clermont-Ferrand, 58, rue Montalembert, 63003 Clermont-Ferrand cedex 1, France*
[b] *Laboratoire d'anatomopathologie, hôpital Gabriel-Montpied, CHU de Clermont-Ferrand, 58, rue Montalembert, 63003 Clermont-Ferrand cedex 1, France*
[c] *Service de radiologie, hôpital Gabriel-Montpied, CHU de Clermont-Ferrand, 58, rue Montalembert, 63003 Clermont-Ferrand cedex 1, France*

Clinical history

A 78-year-old man was referred to ENT for intubation problems encountered before surgery for inguinal hernia. The anesthetist reported difficulty in introducing the intubation tube into the trachea, requiring a change for a smaller caliber. The patient had history of arterial hypertension and prostate surgery. He was free of respiratory symptoms. He was referred to you with an AP pulmonary radiograph (Fig. 1).

Laryngotracheal fibroscopy found an irregular stenosis with nodosities around the entire circumference of the trachea from the subglottis to the carina (Fig. 2).

Cervicothoracic CT (Figs. 3—4), bronchial fibroscopy and biopsy for anatomopathology (Fig. 5) with classic and Congo red staining and a functional respiration test were requested. The biological assessment and functional respiration test results were normal.

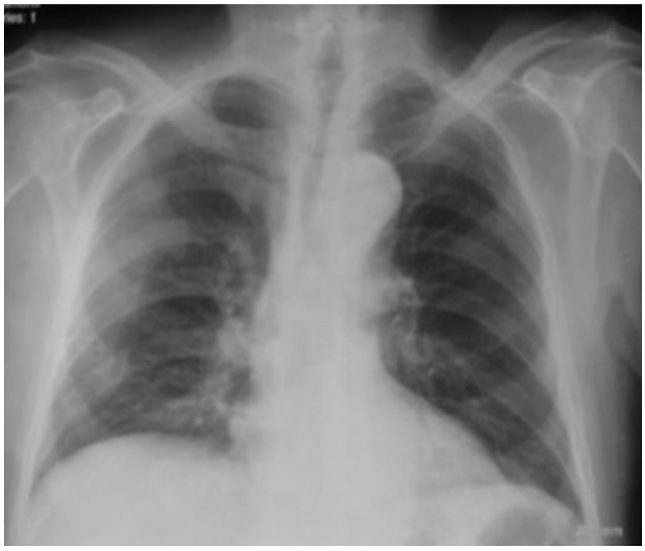

Figure 1 Pulmonary radiograph.

* Corresponding author.
E-mail address: nicolas.saroul@gmail.com (N. Saroul).

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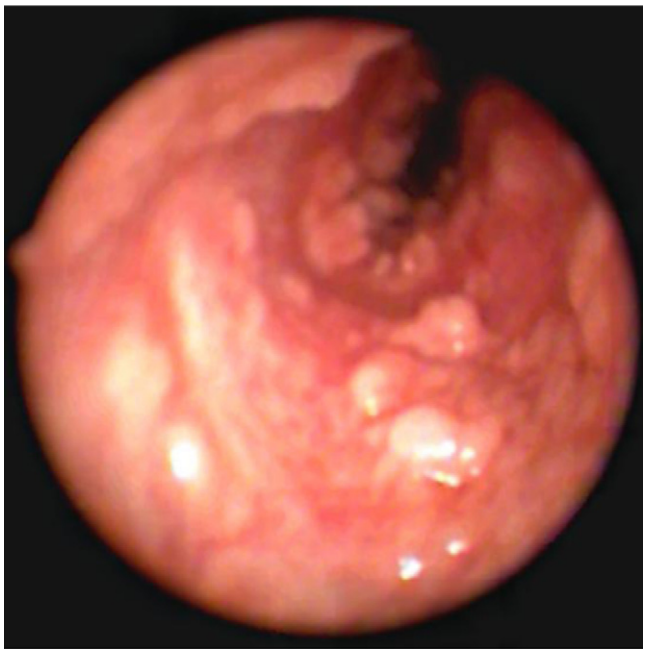

Figure 2 Tracheal fibroscopy.

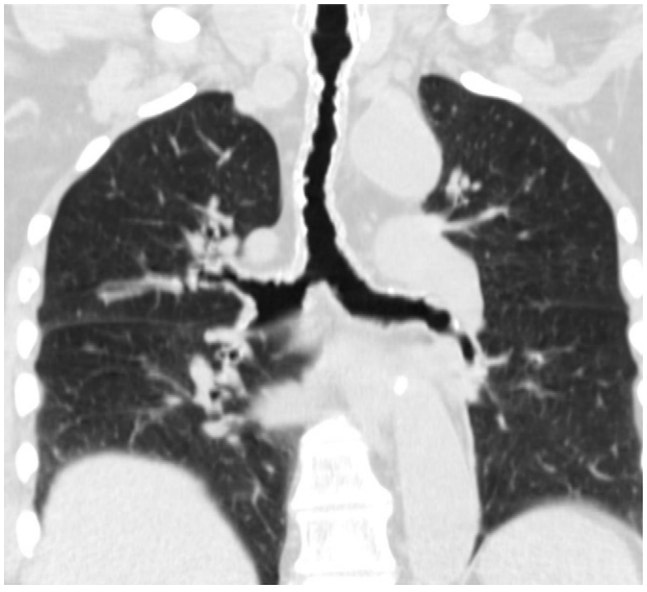

Figure 3 Thoracic CT, coronal slice.

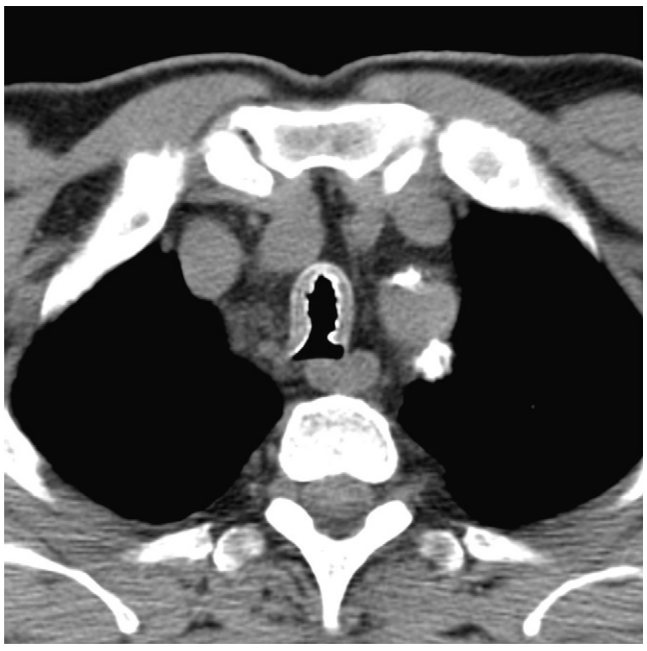

Figure 4 Thoracic CT, transverse slice.

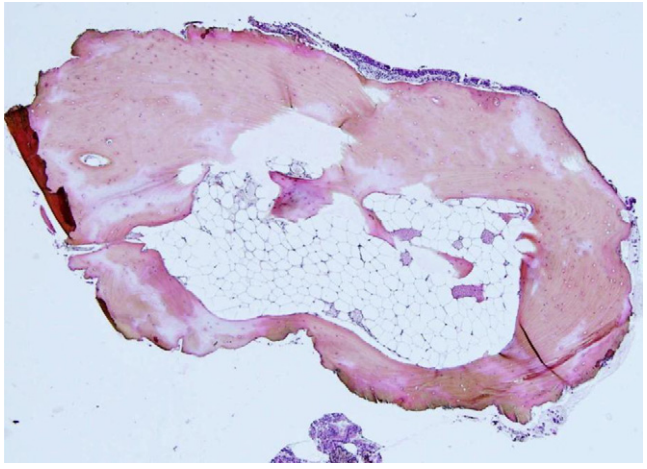

Figure 5 Bronchial biopsy.

Question 1: How do you interpret the images?
Question 2: What is your diagnosis?
Question 3: What treatment do you prescribe?

What is your diagnosis?

Replies

Answer n° 1

Fig. 1 is an AP chest X-ray. Tracheal diameter can be seen to be irregular, with a tendency to stenosis in the inferior part. The pulmonary parenchyma and mediastinum appear normal. Notably, there was no image of extrinsic tracheal compression.

Fig. 2 presents an aspect of tracheal stenosis with nodosities around the entire circumference. The mucosa shows a normal aspect.

Further examination found extension from subglottis to carina.

Figs 3—4 are thoracic CT scans with a parenchymal window. The tracheal aspect was dystrophic, irregular, with multiple nodosities on the anterior and lateral walls. The trachea was calcified, with an undulatory aspect. The posterior wall was lesion-free. The degree of stenosis varied along the trachea. There was no image of extrinsic tracheal compression. The pulmonary parenchyma was normal, and there was no mediastinal adenopathy.

Fig. 5 is the anatomopathologic preparation, with an HEC-stained biopsy sample of bronchial nodosity taken on flexible endoscopy. The bronchial mucosa was well-differentiated with some underlying inflammatory mononuclear elements and an area of osteomedullary tissue comprising compact osseous tissue around an adipocyte-filled medullary cavity. The sample contained no lesions suggesting malignancy.

Answer nº 2

The diagnosis of tracheopathia osteochondroplastica was based on:

- the characteristic aspect of tracheal nodosities with normal mucosa, respecting the posterior wall; that there was no impact on general health status and discovery was serendipitous was also indicative;
- the characteristic CT aspect of a deformed, granular trachea, partially calcified with conserved posterior wall;
- anatomopathologic examination, ruling out differential diagnoses.

Answer nº 3

Simple surveillance was decided on, due to relative lack of evolution and symptoms. At 6 months' follow-up, the aspect was unchanged and symptomatology had not evolved.

Comments

Tracheopathia osteochondroplastica is a rare pathology of unknown etiology involving the large bronchial trunks. Incidence and prevalence are unknown. Onset is adult, with a sex ratio of 1. It is characterized by the development of osseous and cartilaginous nodules under the tracheal mucosa and principal bronchi without extension to the cartilage structures of the respiratory tree [1]. In rare cases, the larynx may also be involved.

Clinical signs are variable and non-specific [1]: chronic cough, recurrent pulmonary infection due to impaired mucociliary clearance in the bronchial epithelium, and recurrent mild hemoptysis. The difficulty lies in recognizing this rare affection before embarking on a series of unproductive complementary examinations. Diagnosis is suggested on CT, and confirmed on endoscopy: an aspect of tracheal nodosities with normal mucosa is pathognomic for tracheopathia osteochondroplastica, and histologic confirmation may be superfluous.

There are three possible differential diagnoses for such cartilaginous airway thickening or stenosis: tracheobronchial amyloidosis, atrophic polychondritis, or a granulomatous pathology such as Wegener's granulomatosis, tuberculosis, sarcoidosis, etc. [2]. The closest diagnosis is tracheobronchial amyloidosis, in which 22% of patients present associated tracheopathia osteochondroplastica [3]; the endoscopic aspect, however, is completely different, featuring grayish, crumbly submucosal plaques and nodules that bleed on contact and extend to the posterior wall of the trachea. In Piazza's series [3], tracheopathia osteochondroplastica was discovered serendipitously on anatomopathologic exploration for suspected amyloidosis at diagnosis or follow-up. Atrophic polychondritis is generally associated with clinical symptoms suggesting chondritis; the tracheal component comprises stenosis without nodosities and sometimes with calcifications that are revealed only on CT [4]. Granulomatous pathologies show various endoscopic aspects (granulation, ulceration, edema, non-specific bronchitis, etc.), but never include the nodosities typical of tracheopathia osteochondroplastica [5].

Tracheopathia osteochondroplastica shows very little evolution over a period of years. In Leske's series [1], there were no directly imputable deaths. Treatment is therefore symptomatic. Non-specific management of intercurrent episodes of infection is important. In a minority of cases, major airway obstruction can develop, requiring local treatment (laser, surgery, dilation [2]). No specific treatment is presently available.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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