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A rare localization of laryngeal sarcoidosis: A case report

Karim Khaoula[1], Bencheikh Razika[2], Benbouzid Mohammed Anas[3], Essakalli Hossyni Leila[4]
[1-4] ENT Department, Rabat Specialty Hospital, Ibn Sina University Hospital, Hafiane-Cherkaoui Avenue, Rabat, Morocco

Abstract
Sarcoidosis is a chronic granulomatous disease affecting people worldwide. The evolution can be acute, with a high rate of spontaneous remission, or chronic evolution, with specific organic manifestations. It affects the upper respiratory tract, of which laryngeal and nasal involvement are the most common.
Laryngeal involvement is most often expressed by dysphonia, dyspnea and chronic cough, which are potentially common symptoms of respiratory failure. it mainly affects the supraglottic region, the diagnosis is based on clinical Biological and radiological findings and supported by histology, which shows noncaseous granulomas. Early diagnosis and adequate treatment are important to prevent airway obstruction, and the use of a tracheotomy. The treatment is based on systemic corticosteroids, immunosuppressants, intra-lesional steroid injection, radiotherapy, CO 2 laser or surgical excision. The evolution of sarcoidosis is unpredictable. It is characterized by relapses and remissions.

Keywords: Dyspnea, granulomatosis, stenosis, tracheaostomy, corticosteroids

1. Introduction
Sarcoidosis is a granulomatous inflammatory disease with predominantly mediastinopulmonary expression, but which can be multi-organ specific, particularly with involvement of the oto-rhino-laryngeal sphere (ENT).
At the laryngeal level, the most commonly involved area is the epiglottis followed by arytenoids, aryepiglottic folds, and ventricular bands.
The involvement of the subglottis is rare.
We report the case of a patient with dyspnoea revealing laryngeal sarcoidosis.

2. Case report
It is a 49-year-old woman born in Mauritania, diabetic on Metformin, who has had a progressive breathing gene for exercise for two years without any other associated sign.
She was admitted to the ENT emergency department for isolated severe laryngeal dyspnea requiring emergency tracheotomy.
The clinical examination was without particularity.
Laryngeal CT scan shows proximal glottic and tracheal stenosis 1 cm long (Figure 1). Direct laryngoscopy: visualization of subglottic stenosis of more than 50% of subglottic lumen at 1 cm in length.
The analysis of two biopsies performed at the level of the stenosis was in favor of a non-specific chronic fibro-inflammatory reshaping, the third biopsy revealed epithelial giant cell granulomatosis without caseous necrosis.
The rhinological examination with nasal endoscopy does not reveal an abnormality of the nasal mucosa.
Thoracic CT scan: non-specific pulmonary micronodule.
Ophthalmological examination: Left iris nodule in favor of sarcoidosis
Salivary gland biopsy: stage 3 lymphocyte sialedenitis
Biological assessment: Sedimentation rate at 53 mm, ECA: 90 UECA, the ANA and p-ANCA are negatives, renal and hepatic assessment was unremarkable.
Syphilitic serology, HIV, hepatitis B and C are negative.
The diagnosis was sarcoidosis laryngeal localization; the patient was put on corticosteroid prednisone at a dose of 1 mg / kg of body weight per day for 6 weeks, followed by a gradual decrease over 6 months.
The clinical evolution was good with a decline of one year.

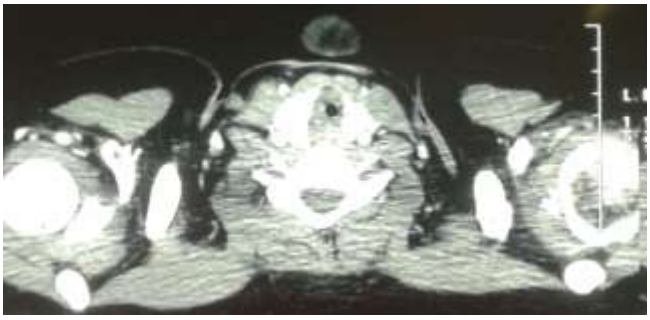

Fig 1: Cervical CT scan showing subglottic circumferential lesion

3. Discussion
Sarcoidosis is a systemic disease whose clinical manifestations can be generalized or localized to one or more organs. It affects the upper respiratory tract in about 6% of cases. The involvement of the head and neck is rare [1].
The nasal involvement is found in 4% of cases, which is mainly manifested by nasal congestion. The disease can also affect the tongue, pharynx, tonsils and nasal bones [2].
The estimated incidence of laryngeal involvement in patients with sarcoidosis is between 1% and 5% [3]. It mainly affects the supraglottic region, less often the subglottis and very rarely the glottal involvement. However, there have been reported cases of paralysis of the vocal cord secondary to peripheral neuropathy by involvement of the laryngeal nerve [4].
The diagnosis is based on clinical, biological and radiological findings and supported by histology, which shows noncaseous granulomas [5].

Laryngeal involvement is most often expressed by dysphonia (63% of cases), dyspnea and cough, found in 47% and 13% of laryngeal cases respectively, are potentially common symptoms of low respiratory involvement. Dysphagia or foreign body sensation is also described [6].

Severe stridors and airway obstruction requiring tracheostomy have been described [7].

Laryngoscopy and tomography of the neck can help evaluate laryngeal structures. Diffuse edema of the supraglottic region is considered the most common manifestation of laryngeal sarcoidosis, and even considered pathognomonic [4].

In our case the lesion was subglottic, considered a very rare localization.

The most commonly involved area is the epiglottis followed by arytenoids, aryepiglottic folds and ventricular bands; sub-glottis involvement is rare, true vocal cords are considered spared due to lack of lymphatic cells [2], only one case has been reported in literature [5].

Due to multisystemic involvement, the patient should have a generalized assessment including pulmonary imaging, hematocrit, liver enzymes, creatinine, ACE, calcemia, including lung imaging, total number of blood cells, liver enzymes, creatinine, ACE, anti-neutrophil cytoplasmic autoantibodies (ANCA), respiratory function testing, electrocardiography and ophthalmological examination.

There are no specific examinations that confirm the diagnosis since ACE is high in 60% of patients with acute illness [8].

Gallium-67 scintigraphy helps diagnosis, although it may be positive for other granulomatosis [9].

Recent publications have demonstrated the validity and utility of PET / CT scan in the diagnosis of sarcoidosis and evaluation of the response to treatment [10].

The histology shows granulomas composed of epithelioid cells, some giant cells, lymphocytes and plasma cells. There is no necrosis or caseation. The search for a granuloma is not specific to sarcoidosis.

The differential diagnosis includes specific infections such as tuberculosis, syphilis, histoplasmosis, coccidiomycosis, actinomycosis, and blastomycosis. Autoimmune diseases such as amyloidosis, Wegener's disease are also included. Malignant neoplasms, cartilaginous tumors, lymphomas, post-traumatic lesions after intubation or laryngotracheal idiopathic stenoses can be evoked [1].

Because of their common histology, and a difficult anatomo-pathological distinction, the suspicion of ENT sites of sarcoidosis must also evoke the existence of upper respiratory tract related to Crohn's disease, despite their extreme rarity.

ENT involvement in Crohn's disease has aspects of oral and gingival ulceration, or edema of the jugular mucosa with pavement appearance [11]. A laryngeal lesion of Crohn's disease is even more exceptional and appears in the form of ulcerations, edema or granulation of the laryngeal mucosa. In the literature, published cases have previously suffered intestinal involvement of Crhon's disease, and other extraintestinal manifestations [12].

Early diagnosis and adequate treatment are important to prevent airway obstruction [4].

The main treatment concerns corticosteroids, while immunosuppressants (methotrexate) or synthetic antimalarials are second-line options. The usual treatment is with prednisone at doses of 1 mg / kg body weight per day for 4 --- 6 weeks, followed by a gradual decrease over 4 --- 6 months [9].

Sometimes laryngeal sarcoidosis may require more aggressive techniques such as corticosteroid injection by laryngeal microsurgery [13].

Endoscopic dilatation or CO2 laser resection [2].

Radiotherapy has been proposed and used in adult patients who do not respond to corticosteroids or when other treatments are not feasible [4].

Finally, tracheotomy may become necessary in cases of severe, recurrent and perennial obstructions. Oral intubation may be an alternative, but it is not always possible [1].

The evolution of sarcoidosis is unpredictable. It is characterized by relapses and remissions and finally "burn out" in 30% of cases, the evolution is more than 5 years considered chronic [13].

4. Conclusion

ENT involvement of sarcoidosis may be in the foreground, and often causes therapeutic problems. The evolutionary profile of ENT lesions and their response to treatment are indeed variable.

5. References

1. Yanardağ H, Enoz M, Papila I, Uygun S, Caner M, Karayel T. Upper respiratory tract involvement of sarcoidosis in the Turkish population. Otolaryngol Head Neck Surg. 2006; 134:848-851.
2. Excurra Hernández P, Albertí I, Casas A, Figuerola Massana E, Blázquez Vilá S, Sempere Durán T *et al*. Sarcoidosis laríngea aislada. A propósito de un caso. Acta Otorrinolaringol Esp. 2003; 54:117-20.
3. Emke MJM. van den Broek. Laryngeal Sarcoidosis: A Case Report Presenting Transglottic Involvement, Depart-ment of Otorhinolaryngology/Head and Neck Surgery, Leiden University Medical Centre, Albinusdreef 2, PO Box 9600, 2300 RC Leiden, The Netherlands, 2013.
4. Dean CM, Sataloff RT, Hawkshaw MJ, Pribitkin E. Laryngeal sarcoidosis. J Voice. 2002; 16:283-8.
5. American Thoracic Society. Statement on sarcoidosis. Am J Respir CritCare Med. 1999; 160:736-755.
6. Bower JS, B Elen JE, WEG JG, D Antzker DR. Manifestations and treatment of laryngeal sarcoidosis. Am Rev Respir Dis. 1980; 122:325-32.
7. Davis C, Girzadas DV Jr. Laryngeal sarcoidosis causing acute upper airway obstruction. Am J Emerg Med. 2008; 26:114, e1–114.e3.
8. Ángel Lede Barreiro A, Juan Díaz Argüello J, José AB. Fernández Martínez, A Alfonso Martínez Ferrerasb Laryngeal Sarcoidosis: Unique Location or First Manifestation Acta Otorrinolaringol Esp. 2012; 63(3):230-232.
9. Baughman RP, Lower EE. Sarcoidosis. In: Fauci AS, Braunwald E, Kasper DL, Hauser SL, Longo DL, Jameson JL *et al*., editors. Harrison. Principios de

Medicina Interna. 17th ed. Available from: www.harrisonmedicina.com/content.aspx?aID=3743621[Chapter 322])
10. Braun JJ, Klesser R, Constantinesco A, Imperiale A. 18FFDG PET/CT in sarcoidosis management: review and report of 20 cases. Eur J Nucl Med Mol Imaging. 2008; 35:1537-43.
11. Arethers CJM, ONNELL MCDWM. Extraintestinal manifestations of Crohn's disease. N Engl J Med. 1994; 330:1870.
12. GIANOLLI GJ, MILLER R. Crohn's disease of the larynx. J Laryngol Otol. 1994; 108:596-8.
13. Mortensen M, Woo P. Office steroid injections of the larynx. Laryngoscope. 2006; 116:1735-9.