CASE STUDY

Laryngeal Sarcoidosis: Unique Location or First Manifestation?☆

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KEYWORDS
Laryngeal sarcoidosis; Systemic granulomatous disorder; Odynophagia

Abstract  Sarcoidosis is a chronic multisystem disease of undetermined aetiology that involves mainly the lung. Affectation of the upper airways is rare, and it is very infrequent as the first manifestation. We report a case of laryngeal sarcoidosis in a 20-year-old woman. The diagnosis was based on clinical information, radiology, histopathology and laboratory tests. The treatment is systemic corticosteroids, as in the pulmonary sarcoidosis. © 2010 Elsevier España, S.L. All rights reserved.

PALABRAS CLAVE
Sarcoidosis laringea; Enfermedad granulomatosa sistémica; Odinofagia

Sarcoidosis laringea: ¿localización única o primera manifestación?

Resumen  La sarcoidosis en una enfermedad crónica multisistémica de causa desconocida que afecta principalmente al pulmón. La afectación de la vía respiratoria superior es rara, e infrecuente como primera manifestación. Presentamos un caso de sarcoidosis laringea en una mujer de 20 años. El diagnóstico se basa la clínica, la radiología, pruebas de laboratorio e histología. El tratamiento, como en la sarcoidosis pulmonar, consiste en corticoterapia sistémica. © 2010 Elsevier España, S.L. Todos los derechos reservados.

Introduction

Sarcoidosis is a multisystemic chronic disease of unknown cause. The course of the disease is chronic, with exacerbations and remissions. It can affect any part of the organism, although it has predilection for the lung. 1 Ocular, spleen, upper respiratory and central nervous system involvement have been reported less frequently. 2

Clinical Case

We report the case of a 20-year old woman with no history of interest, attending consultation due to daily morning odynophagia, without any other symptoms. Laryngoscopy showed an enlargement of the left arytenoepiglottic band and fold, with hyperaemic and regular mucosa.

Chest radiography showed no pleuropulmonary abnormalities. A cervical CT scan showed hyper-uptake by a mass measuring 2×1.5 cm in the left arytenoepiglottic fold (Fig. 1A). An MRI scan showed focal thickening of the left arytenoepiglottic fold, with diffuse contrast uptake, accompanied by segmental uptakes in both bands (Fig. 1B). Flexible oesophagoscopy was normal. A general analysis was performed, which revealed the following values: angiotensin


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Figure 1  (A) CT scan: hyper-uptake by a mass in the left arytenoepiglottic fold. (B) MRI scan: thickening of the left arytenoepiglottic fold, with diffuse contrast uptake.

Sarcoidosis affects the larynx in up to 5% of cases, predominantly in the supraglottic region (especially the epiglottis). Subglottic involvement is less frequent and glottic cases are even rarer. However, there have been reports of vocal cord paralysis secondary to peripheral neuropathy due to recurrent laryngeal nerve involvement.

Diagnosis is based on the symptoms, ENT examination, radiographic findings, laboratory tests and histology. Symptoms can be nonspecific, predominantly hoarseness, dyspnoea and dysphagia and, less frequently, foreign body sensation, wheezing and cough. Occasionally, it can reach laryngeal stridor and airway obstruction.

Discussion

Sarcoidosis is a systemic disease whose clinical manifestations can be generalised or localised in one or more organs. Almost always there is pulmonary involvement and respiratory symptoms. Between 2% and 15% of systemic cases sarcoidosis present otolaryngological manifestations. Nasal involvement is found in up to 4% of cases, appearing as nasal congestion. The disease may also affect the tongue, pharynx, tonsils and nasal bones.

Poe was the first to describe a case of laryngeal involvement in 1940. Isolated laryngeal sarcoidosis with no evidence of systemic disease is rare, which led us to conduct long term monitoring.

converting enzyme (ACE) (23.3 IU/l), ESR (33.0 mm/h), C reactive protein (15.2 mg/l), rheumatoid factor (<10.0), ANA and anti-DNA (negative).

We performed biopsy (laryngeal microsurgery), which was reported as non-necrotising granulomatosus inflammation with epithelioid cells (Ziehl–Neelsen staining was negative), compatible with sarcoidosis, as well as negative Lowenstein culture.

With this diagnosis, we initiated treated with systemic glucocorticoids, which rapidly improved the symptoms. At 6 months, laryngoscopy was normal.

At 2 years, the patient consulted again due to odynophagia and pharyngeal foreign body sensation. Laryngoscopy showed a small tumour with normal mucosa on the right arytenoepiglottic fold and ventricular band, with oedema in the retrocricoid region and pyriform sinuses (Fig. 2). A CT scan showed a circumferential thickening of soft tissue which obliterated the pyriform sinuses and affected the arytenoepiglottic folds, bands and vocal cords. The length of the affected segment was approximately 3.3 cm. A new steroid treatment improved the symptoms within 1 month, and exploration was normal at 6 months.

Figure 2  Laryngoscopy (after reactivation of symptoms): tumour mass on right arytenoepiglottic fold and ventricular band.
An oedematous, pale and diffuse increase of the supraglottic area is regarded as the most common manifestation of laryngeal sarcoidosis, and even considered pathognomonic.\textsuperscript{5,7}

There are no laboratory studies which confirm the diagnosis, since ACE is elevated in 60% of patients with acute illness. The Kviem–Siltzbach test is a specific test, but it is discarded due to its technical difficulty.\textsuperscript{1,5} Gallium-67 scintigraphy supports the diagnosis, although it also shows a common pattern with other granulomatous diseases.\textsuperscript{1,5} Recent publications have demonstrated the validity and usefulness of PET/CT scans in the diagnosis of sarcoidosis\textsuperscript{8} and the evaluation of response to treatment.\textsuperscript{9}

Establishing the diagnosis of isolated laryngeal sarcoidosis involves demonstrating a past and present absence of systematisation of the disease\textsuperscript{3} or, as already mentioned, in the future. Thus, constant monitoring of these patients is crucial. Inflammatory diseases such as Wegener’s granulomatosis, amyloidosis, rheumatoid arthritis or systemic lupus erythematosus can affect the larynx, and display similar symptoms, histopathology and images.\textsuperscript{6}

The prognosis of sarcoidosis is good. Most patients with acute disease recover without relevant sequelae.\textsuperscript{1} There have been reports of spontaneous regression.\textsuperscript{6}

Early diagnosis and adequate treatment are important to prevent airway obstruction.\textsuperscript{2} The main treatment is with corticosteroids, while immunosuppressants (methotrexate) or 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, subsequently followed by a gradual decrease over 4–6 months. If the disease recurs, the cycle is repeated.\textsuperscript{1} Sometimes, laryngeal sarcoidosis may require more aggressive techniques such as intraleisonal corticosteroid injection using laryngeal microsurgery,\textsuperscript{10} endoscopic dilation or CO\textsubscript{2} laser resection.\textsuperscript{2} Radiotherapy has been proposed and used in adult patients unresponsive to corticosteroids or when other treatments are not feasible.\textsuperscript{5,11}

Finally, tracheotomy may become necessary in cases of severe, recurrent, life-threatening obstructions. Oral intubation may be an alternative, but it is not always possible.\textsuperscript{12}

Conflict of Interests

The authors have no conflicts of interest to declare.

References