CASE STUDY

Sarcoidosis in the Nasopharynx, a Rare Location

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Abstract

Sarcoidosis is a systemic granulomatous disease that usually has a pulmonary presentation. The extrapulmonary organs most frequently affected are lymph nodes, eyes and skin. Rhinopharyngeal involvement is extremely rare. We describe a case of sarcoidosis which was diagnosed through its location in cavum.

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Clinical Case

We present the case of a 45-year-old woman who attended consultation due to bilateral nasal respiratory insufficiency—rhinorrhoea and bilateral hypoacusis of one year's evolution. In addition, she suffered episodes of asthenia, fever and pretibial erythema nodosum on both legs.

Otoscopy revealed bilateral serous otitis. Nasal fibroscopy enabled us to visualise a tumour in the nasopharynx, obstructing both choanae. The rest of the ENT examination was normal. The MRI revealed a soft tissue mass in the nasopharynx; the mass was obstructive and compatible with adenoid hypertrophy (Fig. 1).

We performed an endoscopic biopsy of the tumour under local anaesthesia with the following pathological report: lymphoid tissue in which it was possible to identify numerous small non-necrotizing granulomas composed of epithelioid histiocytes and multinucleated giant cells.

Given these findings we began a differential study. Staining techniques of the sample (Ziehl-Nielsen, PAS, silver) ruled out microorganisms. The serological tests for brucella, lues and HIV, as well as Mantoux, resulted negative. The analytical study showed only an elevation in ESR and ACE (angiotensin-converting enzyme). The remaining tests—blood count, biochemistry, rheumatoid factor, antinuclear antibodies (ANA) and serum protein—were normal.

PALABRAS CLAVE

Sarcoidosis; Nasofaringe; Vía respiratoria superior

Sarcoidosis en nasofaringe, una extraña localización

Resumen

La sarcoidosis es una enfermedad granulomatosa sistémica que generalmente afecta al pulmón. Las localizaciones extrapulmonares más frecuentes son los ganglios linfáticos, los ojos y la piel, mientras que en la rinofaringe es extremadamente rara. Presentamos un caso de sarcoidosis que fue diagnosticada por su localización en el cavum.

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Chest radiography showed no pathological findings. The patient was referred to the internal medicine service, which ruled out systemic involvement.

In view of the result of non-necrotizing granulomas in the nasopharynx, episodes of asthenia, erythema nodosum and ACE elevation and after ruling out other diseases, we diagnosed sarcoidosis in the nasopharynx, with no other companion initial locations.

Given that this was a localized form of sarcoidosis without serious impact, we decided to initiate conservative treatment with topical mometasone 1 inh/24 h. In 3 months, the mass had decreased in size and the respiratory insufficiency and seromucinous otitis media had disappeared (Fig. 2).

Significant growth took place after 8 months and, with it, the symptoms reappeared. Because the topical corticosteroid regime failed, we started treatment with oral deflazacort at doses of 1 mg/(kg day) in a descending pattern for 12 days. Subsequent reviews showed that the tumour decreased gradually and then disappeared. Follow-up after one year found no recurrence or other systemic disease.

Discussion

Sarcoidosis is a chronic, systemic disease, whose aetiology and pathogenesis are unknown. Spain has an incidence of 0.05–1.2 cases/100,000 population/year. It affects the ENT area in 2%–15% of cases. It has been described in the lymph nodes, parotid, larynx and nasal fossae, among other locations. Its location in the nasopharynx is extremely rare and there are very few cases in the literature. The diagnosis requires: compatible clinical findings, histological confirmation of non-caseating epithelioid granulomas and the exclusion of other pathologies (mycobacteria, syphilis, fungi, neoplasms and Wegener granulomatosis). The disease remits spontaneously in most cases, although it occasionally progresses towards chronicity. Consequently, treatment would be indicated for only those cases where there is potential risk or progression of the symptoms. In general, corticosteroids are the treatment of choice, at a dose of 0.5 mg/(kg day), attempting to reduce this initial dose to the lowest effective one. Localised nasal forms can be treated with topical corticosteroids; these may even be used in the form of intralesional injections. Surgery may be an alternative in circumscribed or obstructive cases, or those resistant to medical treatment.

In conclusion, although the nasopharynx is a rare location for sarcoidosis, we believe that it should be considered in the differential diagnosis of tumours of the nasopharynx. This is a suspicion and exclusion diagnosis. Treatment should initially be conservative and always individualised.

References