Case Report

Isolated sinonasal sarcoidosis with intracranial extension: case report

Sarcoidosi isolata sinusonasale con estensione intracranica: caso clinico

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SUMMARY

The case is presented of a 20-year-old female who was incidentally diagnosed with isolated sarcoidosis in the form of a nasal mass involving the ethmoid sinuses with destruction of the cribriform plate and intracranial extension. The patient complained of cacosmia and nasal obstruction. Histopathologic examination of biopsies from the nasal mass led to a diagnosis of non-caseating granuloma, highly suggestive of sarcoidosis. No systemic manifestations were detected, and the patient was started on a regimen of systemic steroids. Over the 3 months following biopsy and commencement of systemic steroids, the patient showed improvement in symptoms, and computed tomography of the paranasal sinuses revealed complete resolution of the sinonasal mass. Follow-up for one year revealed no recurrence.

KEY WORDS: Nose • Paranasal sinuses • Granulomatous disease • Sarcoidosis • Corticosteroid therapy

RIASSUNTO

Presentiamo il caso di una donna di 20 anni, in cui è stata diagnosticata incidentalmente una sarcoidosi isolata nasale con coinvolgimento del seno etmoidale, distruzione della lamina cribra ed estensione intracranica. La paziente lamentava cacosmia e ostruzione nasale. L'esame istopatologico della biopsia eseguita sulla massa nasale ha evidenziato un granuloma non caseoso, altamente indicativo di una sarcoidosi. Nessuna manifestazione sistemica è stata rilevata e la paziente è stata sottoposta ad un trattamento a base di corticosteroidi per via sistemica. Tre mesi dopo l'inizio della terapia, la paziente riferiva netto miglioramento della sintomatologia e la tomografia computerizzata dei seni paranasali evidenziava una regressione completa della massa sinusonasale. Non è stata evidenziata nessuna recidiva nel corso del primo anno di follow-up.

PAROLE CHIAVE: *Naso • Seni paranasali • Malattia granulomatosa • Sarcoidosi • Terapia cortisonica*

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Introduction

Sarcoidosis is described as a multi-system granulomatous disorder of unknown cause, which, most commonly affects young and middle-aged females and may involve a variety of sites, mainly the lungs, skin, liver, eyes, spleen, peripheral lymph nodes and neural structures [1]. Sinonasal involvement, in the absence of pulmonary disease, is extremely rare [2]. The otorhinolaryngologic signs and symptoms of sarcoidosis are not specific and can mimic other more common disorders [3]. The characteristic lesion of sarcoidosis is the noncaseating granuloma found with tissue biopsy. This can be present in any organ system or areas of the body [4]. Histologically, affected tissues exhibit multiple epithelioid cell granulomas that are organized collections of mature mononuclear cells. Hyaline fibrosis, leukocyte infiltration, necrosis, and refractile structures within the epithelioid cells may also be present. The epithelioid cells secrete cytokines and mediators, including angiotensin converting enzyme (ACE) [5]. This sign is suggestive of sarcoidosis but is not a diagnostic biochemical marker. Further diagnostic tests are usually necessary to exclude other granulomatous disorders, such as tuberculosis, aspergillosis, actinomycosis, Wegener's granulomatosis, Churg-Strauss syndrome and lymphoma [6].

Case report

A 20-year-old female presented with nasal obstruction, cacosmia and hyposmia. The symptoms had been increasing in severity over a 6-month period. The patient's complaints started 18 months earlier, when she noticed a swelling in the outer quadrant of her left orbit. The patient had no ocular or nasal symptoms. Computed tomography (CT) of the paranasal sinuses, orbit, and chest (with contrast) were all normal except for lacrimal gland enlargement (Fig. 1). Six months later, she started developing increasing nasal obstruction, hyposmia progressing to cacosmia. Biopsy of the lacrimal gland was performed which revealed lacrimal gland hyperplasia. Nasal endoscopy revealed a grayish friable mass high up in the roof of the nose on both sides of the nasal septum. Contrast CT of the paranasal sinuses and

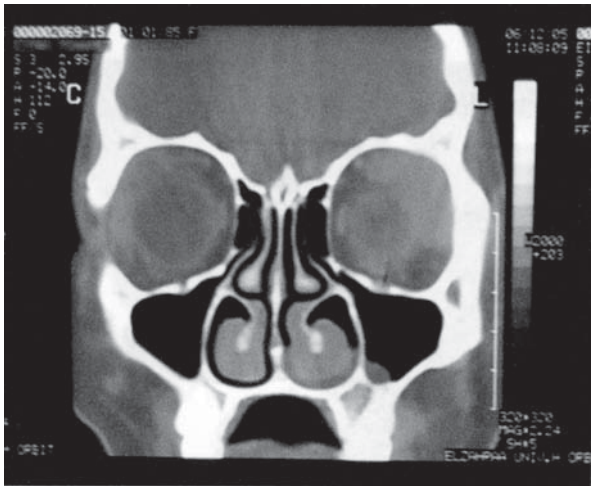

Fig. 1. CT PNS showing left lacrimal gland enlargement, free ethmoid sinuses and nasal roof. NB enlarged inferior turbinates.

magnetic resonance imaging (MRI) of the paranasal sinuses and brain were performed. CT of the paranasal sinuses showed a soft tissue mass occupying the roof of the nose, involving both ethmoid sinuses with erosion of the cribriform plate. The inferior turbinates showed marked enlargement (Fig. 2). The mass was isointense on MRI T1 weighted images, with marked enhancement with gadolinium contrast, hyperintense on T2 weighted images, breaching the cribriform plate with involvement of the lower part of the right frontal lobe of the brain (Fig. 3). The patient had no neurological symptoms and was scheduled for a nasal biopsy. Biopsy was taken from the mass in the roof of the nose and from the infero-lateral aspect of both inferior turbinates. The mass in the roof revealed sheets of inflammatory cells comprising lymphocytes and plasma cells widely separated by non-caseating granulomas composed of central epithelioid cells with multinucleated giant cells (Fig. 4). This was highly suggestive of sarcoidosis. There was no evidence of malignancy. The biopsies from the inferior turbinates were not conclusive. The patient had no systemic manifestations of sarcoidosis and, furthermore, tested negative for Ziehl Nielsen, anti-neutrophil cytoplasmatic antibodies (ANCA), and had normal serum levels of ACE and normal serum

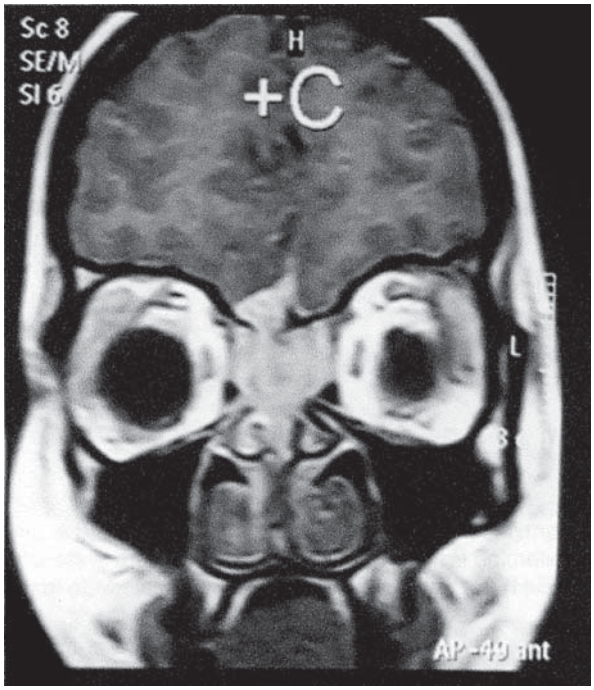

Fig. 3. MRI PNS showing sinonasal mass exhibiting marked enhancement on T1 weighted images with gadolinium contrast. Evident breach of cribriform plate with involvement of lower part of right frontal lobe of brain can be seen.

levels of calcium. Complete blood count, kidney function tests and urine analysis were within the normal range. The patient was put on a course of oral corticosteroids, starting with 60 mg/d prednisolone for 2 weeks, and lowering the dose by 10 mg/d every 2 weeks till 40 mg/d, to be lowered then by 5 mg/d every 2 weeks. After 18 weeks, the patient was maintained on 10 mg/d for another 8 weeks (total = 26 weeks\6 months). This was coupled with nasal cortisone spray (Fluticasone Propionate, twice daily) throughout treatment and a proton pump inhibitor (PPI) (Omeprazole 20 mg, once daily) for the first 2 months. Weekly follow-up included nasal endoscopy, which after 4 weeks of medical treatment showed a gradual involution of the nasal mass. Clinically, the patient felt the sensation of a decrease in

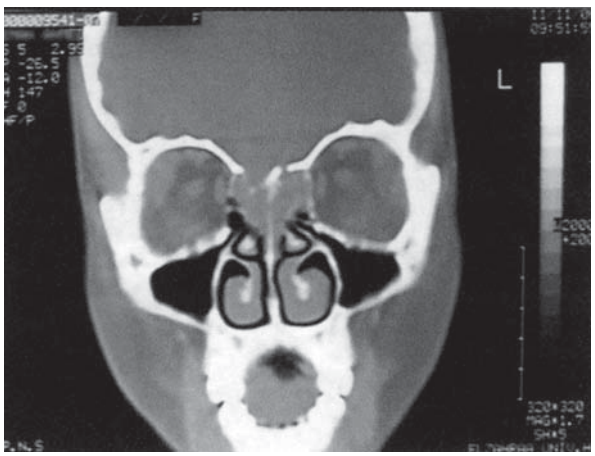

Fig. 2. CT PNS showing sinonasal mass occupying roof of nose and both ethmoidal sinuses. NB destruction of cribriform plate, more on right side.

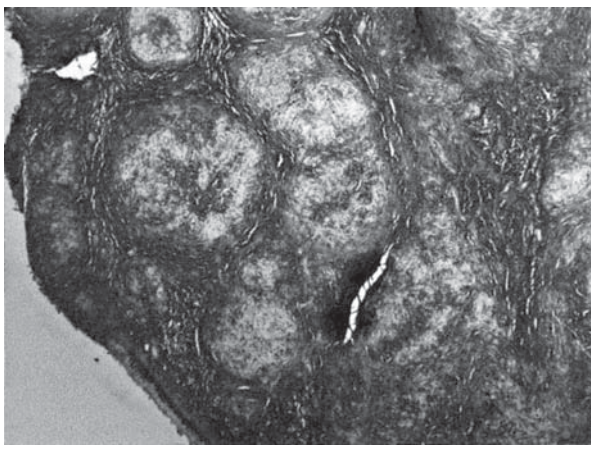

Fig. 4. Sheets of inflammatory cells comprising lymphocytes and plasma cells widely separated by non-caseating granulomas composed of central epithelioid cells with multinucleated giant cells. Respiratory nasal epithelium can be seen on lower left (H&E x40).

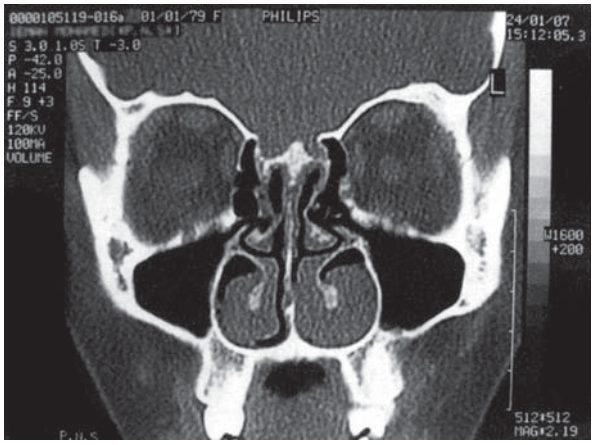

Fig. 5. CT PNS showing complete involution of sinonasal mass.

cacosmia, but still complained of hyposmia. Following 12 weeks of medical treatment, contrast CT of the paranasal sinuses showed complete involution of the nasal mass (Fig. 5). The patient stopped complaining of cacosmia. Endoscopic follow-up was performed monthly. Six months after commencing treatment, follow-up CT of the paranasal sinuses confirmed the absence of any sinonasal lesion. Medication was stopped, and the patient was observed at follow-up for a further 6 months with endoscopic examination. No recurrence was found.

Discussion

Sarcoidosis, principally affecting the respiratory tract, rarely involves the head and neck region and can be a diagnostic challenge for the otolaryngologist. Granulomatous involvement of nasal mucosa, already described by Boeck in 1905, is, however, rare [6].

In patients with sarcoidosis, sinonasal involvement may develop, but the disease is rarely isolated to this area [7]. McCaffrey and McDonald [8] reviewed the records of 2319 patients diagnosed with sarcoidosis and found nasal mucosa involvement in 17 (less than 1%). Wilson et al. [7] noted nasal involvement, confirmed by biopsy, in 21 (2.8%) out of 750 patients with sarcoidosis. A recent report by Zeitlin et al. [9] stated a 4% incidence of nasal involvement in 159 patients with sarcoidosis. Those Authors also noted that the actual incidence might be much higher. The most frequent sites of nasal involvement are the nasal septum and inferior turbinates, followed by the paranasal sinuses, nasal bone and cartilage, and subcutaneous tissues in the region [10]. The prognosis of patients with sarcoidosis is generally good, however mortality rates are reported to vary from 5-7% [11]. There is no mention, in the otolaryngologic literature, to the best of our knowledge, of isolated sinonasal sarcoidosis with intracranial extension. Our patient had no lung, skin, liver or any systemic affection. Corticosteroids remain the cornerstone of therapy. Nasal topical steroid application can help control the progression of isolated nasal involvement in certain cases [1], however treatment with systemic steroid is usually needed. Surgery should be the last resort. Patients should be followed carefully over the long term since there is a tendency for recurrence and delayed systemic involvement.

Conclusions

Nasal involvement in sarcoidosis is rare, but otolaryngologists should consider this condition in the differential diagnosis of sinonasal complaints since signs and symptoms of sarcoidosis are not specific and can mimic other more common disorders. Further diagnostic tests are usually necessary to exclude other granulomatous disorders. Corticosteroids remain the cornerstone of therapy. Despite often long and aggressive treatment, relapses and chronicity are frequent after tapering or discontinuing the corticosteroids and require a long follow-up and interdisciplinary management. Further studies are indicated to determine the effectiveness of the different treatments, to evaluate the prognostic factors, and to access the natural history of sinonasal sarcoidosis.

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