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Ethmoidal sarcoidosis

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Abstract
Sarcoidosis is a multisystem granulomatous disease of unknown aetiology. Nasal sarcoidosis commonly affects the mucosa of the septum and the inferior turbinates. These patients may present with nasal discharge, crusting, obstruction, epistaxis or anosmia. We present an unusual case of nasal sarcoidosis involving the ethmoid sinus causing recurrent eyelid swelling and discuss its management.

Key words: Sarcoidosis; Ethmoid sinus; Nose

Introduction

Sarcoidosis is a chronic systemic disease of unknown aetiology. It most commonly affects the lungs, lymph nodes, liver, spleen, eyes and bones. Histopathologically it is characterized by non-caseating granulomatous inflammation of the organs involved. Sarcoidosis of the mucous membrane of the upper respiratory tract is comparatively rare. About one per cent of patients with sarcoidosis have histologically proven nasal mucosal involvement.

We report a case of nasal sarcoidosis, affecting the ethmoid sinuses primarily, which presented with recurrent eyelid swelling and decreased visual acuity. The management of nasal sarcoidosis is discussed.

Case report

In 1985, a 31-year-old man complained of nasal obstruction and discharge. Physical examination revealed bilateral nasal polyps but the remainder of his upper respiratory tract was normal. The skin prick allergy test was negative. Paranasal sinus X-ray films revealed mucosal thickening of all the sinuses. He underwent a nasal polypectomy and the histology showed benign inflammatory polyps. Later that year, he was referred to a respiratory physician complaining of malaise, intermittent wheeze, and cough. Physical examination revealed no abnormalities. A chest X-ray film showed bilateral hilar lymphadenopathy. The Kveim-Siltzbach skin test was positive and biopsy of the mediastinal lymph nodes established the diagnosis of sarcoidosis.

In 1987, he presented to the ophthalmologist with a painful left palpebral swelling and decreased visual acuity affecting the left eye. A CT scan of his orbits showed no orbital lesion except for some enlargement of the left optic nerve. There was diffuse mucosal thickening of all the paranasal sinuses. Biopsy specimens of the lacrimal gland and subconjunctiva showed eosinophil polymorph infiltration but no granulomata were identified. His clinical condition improved completely on a course of systemic steroid therapy.

The patient presented again to the ophthalmologist in March 1992 with similar symptoms affecting the left eye. On examination there was swelling of the left eyelids and a visual acuity of 6/9. There was no proptosis and fundus examination was normal. There was no abnormality in the right eye. A CT scan of his orbits showed no abnormality in either orbits. However, an opacity was present in the left anterior ethmoid sinus (Figure 1). The patient was started on systemic corticosteroid therapy and the visual acuity in his left eye improved to 6/4. He was later referred to the Otolaryngology Department for a further opinion. Intranasal examination showed bilateral nasal polyps and nasal mucosa appeared normal. The rest of the upper respiratory tract was within normal limits. It was decided that the left ethmoid sinus should be explored.

The patient was admitted for left external ethmoidectomy. At operation a polypoidal mucosa was found in the anterior ethmoid cells. Histology of the left ethmoid sinus mucosa revealed a confluent mass of non-caseating epithelioid granulomas consistent with sarcoidosis (Figures 2 and 3). There was an inflammatory infiltrate consisting of plasma cells and lymphocytes. Stains for acid-fast organisms and fungi were negative. Post-operatively he made a good recovery and was treated with topical nasal steroid spray. He has been followed-up in the clinic and to date there has been no recurrence of the visual symptoms in the left eye. Similarly there has been no recurrence of his nasal polyps.

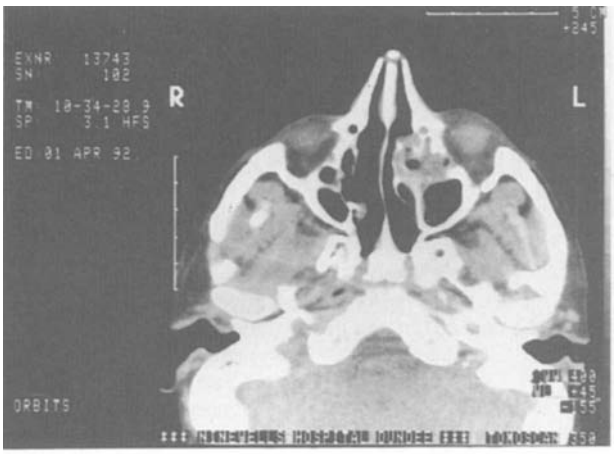

Fig. 1
Pre-operative axial CT scan showing soft tissue mass in the left ethmoid sinus.

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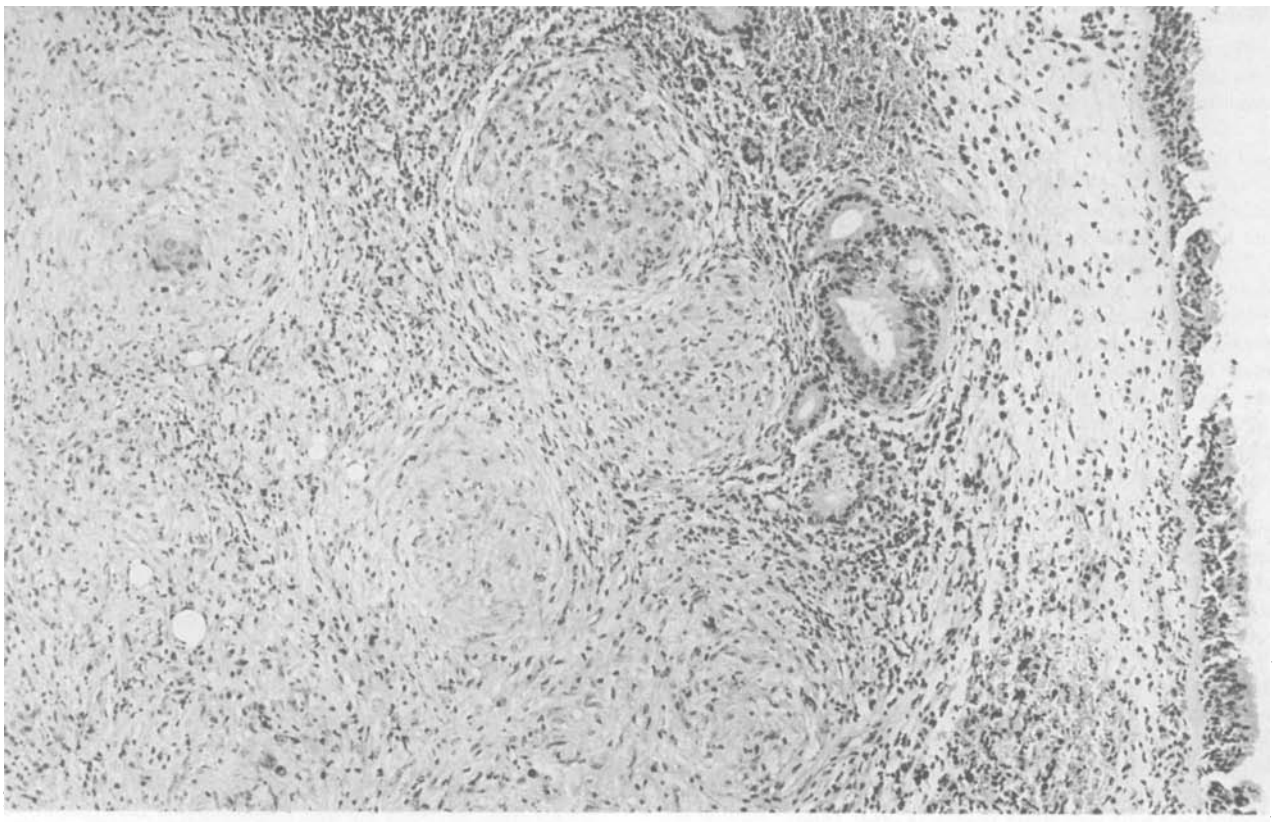

FIG. 2
Photomicrograph showing a confluent mass of non-caseating epithelioid granulomas in the submucosa. (H & E; × 100).

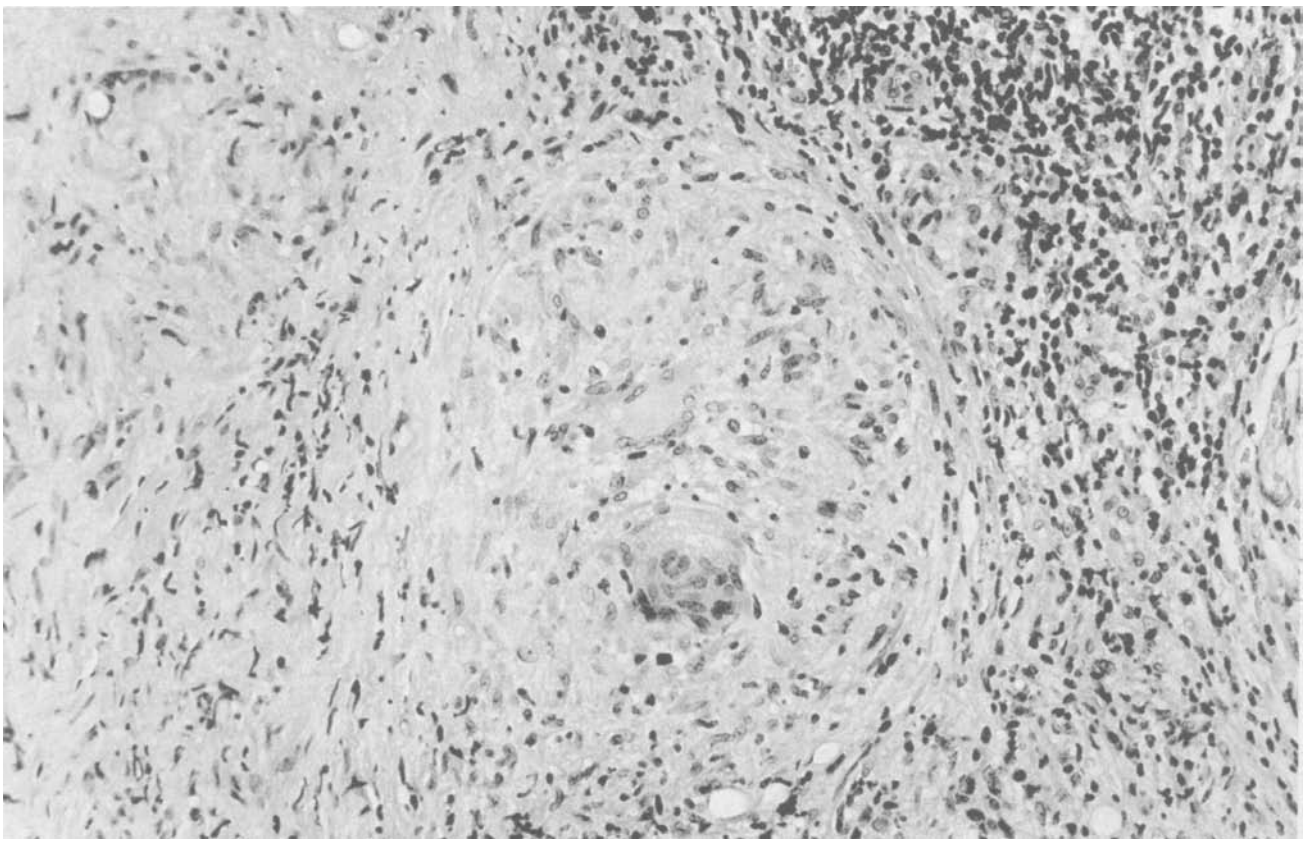

FIG. 3
High power photomicrograph showing epithelioid cells and giant cells in a granuloma. (H & E; × 250).

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Discussion

The incidence of nasal sarcoidosis (Gordon *et al.*, 1976) has been said to vary from three to 20 per cent of cases with systemic sarcoidosis (Lindsay and Perlman, 1951; Weiss, 1960; Neville *et al.*, 1976). However, McCaffrey and McDonald (1983) examined the records of 2319 patients with the diagnosis of sacroidosis at the Mayo Clinic over a 31-year period and found the incidence of nasal involvement to approximate one per cent. It is rare for sarcoidosis to affect the nose exclusively.

The most frequent symptoms related to the nose are nasal obstruction and epistaxis. Other symptoms include a nasal mass lesion, nasal pain and anosmia. Our case was unusual in two respects, i.e. its presentation and its principal site of involvement. To our knowledge, this is the first reported case of recurrent eyelid swelling, secondary to sarcoidosis affecting the ethmoid sinus primarily, without orbital involvement. It also shows that sarcoidosis can affect the paranasal sinuses without any macroscopic changes in the nasal mucosa.

Nasal mucosal lesions usually occur on the anterior septum and inferior turbinates. The mucosa is usually dry and crusting occurs. Occasionally hypertrophic rhinitis with polypoid mucosal changes are seen. The characteristic appearance consists of pale, yellow submucosal nodules, varying from 1 to 7 mm in size. These nodules are believed to be submucosal granulomas. The pathological process may resolve or proceed to fibrosis or atrophic rhinitis. Septal perforation may result from atrophic rhinitis or it may complicate nasal septal surgery especially submucosal resection (Fletcher, 1944; Neville *et al.*, 1976). Lupus pernio is a chronic, violaceous skin lesion of the nose, cheeks and ears. Sarcoidosis may also affect the nasal bone producing osteoporosis with punch-out lesions (Curtis, 1964).

The diagnosis of nasal sarcoidosis is based on the clinical features and the histopathological appearance of non-caseating granulomas. The mucosa of the anterior septum and inferior turbinates provide an easily accessible place for biopsy (Weiss, 1960). However, there is no single investigation that is specifically diagnostic for sarcoidosis and other causes of granulomatous inflammation of the nasal mucosa must be excluded. Certain laboratory findings that may help in the diagnosis include hypercalcaemia and hypercalciuria, an elevated serum angiotensin converting enzyme level, and a positive Kveim-Siltzbach skin test. There is usually an anergy to the tuberculin skin test.

Other granulomatous diseases may affect the nasal mucosa (McCaffrey and McDonald, 1983). Nasal disease may be associated with systemic granulomatous diseases such as Wegener's granulomatosis, polymorphic reticulosis and Churg-Strauss syndrome. Infectious rhinitis with granuloma formation occurs in tuberculosis, actinomycosis, aspergillosis, blastomycosis, histoplasmosis, leprosy, syphilis and rhinoscleroma. Berylliosis can produce foreign body granulomas similar to sarcoidosis but a history of occupational exposure will establish the diagnosis.

The treatment of sarcoidosis will depend on the specific organs involved. The main indication for systemic corticosteroid therapy is when vital organs are threatened and in hypercalcaemia. If the symptoms are confined to the nasal mucosa, topical preparations of corticosteroids have much to offer. There have been reports of spontaneous regression of nasal sarcoidosis (Page and Seth, 1969). Surgery, apart from obtaining biopsy tissues, has not been found to be helpful.

Acknowledgement

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