

CASE REPORT: A RARE CAUSE OF PROPTOSIS

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INTRODUCTION

Proptosis is the forward protrusion of the eye from its normal position in the orbit. There are many causes, the commonest of both unilateral and bilateral proptosis being thyrotoxicosis. Antral tumours are an uncommon cause of proptosis but one that should be considered in any patient with proptosis. A mucocele is a cystic expansile lesion that is lined with epithelium and filled with mucoid secretions⁽¹⁾. It results from disrupted or obstructed drainage of the paranasal sinuses⁽²⁾. Maxillary sinus mucoceles are infrequent and can result in proptosis, nasal obstruction, facial swelling and involvement of the oral cavity.

Primary lymphomas of the sinonasal tract are rare, representing approximately 8% of sinonasal malignancies, the maxillary sinus being the commonest site⁽³⁾. Malignancy is a diagnosis that must be considered in the aetiology of maxillary mucocele and we believe that this is the first documented case of bilateral maxillary mucocele caused by non-Hodgkin's lymphoma.

CASE REPORT

A 74-year-old gentleman was referred to the ENT department with a one-month history of left-sided facial discomfort, left infra-orbital nerve numbness and diplopia on looking left. There was no history of nasal obstruction, discharge, epistaxis or anosmia. Examination revealed left-sided proptosis and confirmed the left-sided infra-orbital weakness. There was no cervical lymphadenopathy.

A CT scan of his sinuses showed a solid homogenous mass involving both maxillary sinuses extending to the anterior ethmoids and orbits (Figure 1). The roofs of both maxillary antra were eroded. The appearances were felt to be suggestive of either infection, mucocele or lymphoma.

An urgent antral lavage was performed under local anaesthetic to relieve the patient's discomfort. The return was a thick creamy pus, which was sent to microbiology and the patient was started on intravenous antibiotics. Following this, the patient underwent bilateral radical antrostomies and intranasal ethmoidectomies under general anaesthesia. The entire sinuses appeared to be full of an infected mucocele and this, along with all the unhealthy-looking mucosa, was removed and sent for histological examination.

Histology revealed antral mucosa infiltrated with atypical lymphocytes which on immunoperoxidase staining revealed these cells to be of B cell origin. The appearances were of diffuse high grade B-cell non-Hodgkin's lymphoma. Following surgery the patient has begun chemotherapy, and at present (eight months post-surgery) remains free from systemic disease and local recurrence.

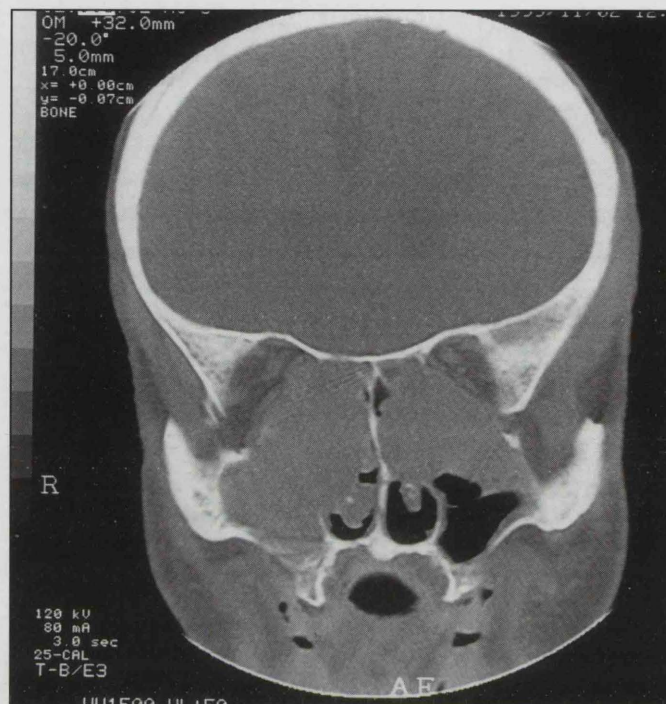


Figure 1 CT showing involvement of both maxillary sinuses

DISCUSSION

The majority of paranasal mucoceles occur in the frontal sinuses (two-thirds) and the ethmoidal sinuses (one-third). Maxillary mucoceles are said to be extremely rare. Obstruction of the ostium in sinus mucoceles may be the result of infection, trauma, previous surgery, tumour, cystic degeneration of the mucosa or mucociliary dysfunction. Continued secretion of the sinus mucosa leads to the air-filled cavity being replaced with mucus. This in turn progresses to expansion of the sinus and thinning of the bone walls, which, if left, can lead to pressure necrosis⁽⁴⁾. Clinically the mucoceles can result in proptosis, diplopia, nasal obstruction, facial swelling and involvement of the oral cavity.

Radiologically, mucoceles appear as clouded sinuses with some expansion of the sinus walls. Only five percent demonstrate macrocalcifications⁽⁵⁾. Appearances on CT often demonstrate the bony erosion clearly⁽⁶⁾ and have been well documented by Som and Shugar⁽⁷⁾. It has been emphasised that early diagnosis can be facilitated by the appropriate use of a CT scan⁽⁸⁾.

Lymphomas of the sinonasal tract are especially rare in Western populations. Of 33,402 cases of nodal and extranodal malignant lymphomas in the Kiel Lymph Node Registry from 1972 to 1987, only 0.17% were in the nasal cavity and paranasal sinuses⁽⁹⁾, and of these only 15% were primarily in the paranasal sinuses⁽¹⁰⁾.

Distinguishing primary involvement of the nasal cavity from primary involvement of the paranasal sinuses has histopathologic and therapeutic implications⁽¹¹⁾.

Sinonasal lymphomas of B-cell phenotype predominate in Western populations⁽¹²⁾. The B cell lymphomas are suspected to derive from precursor cells located in subepithelial or interglandular lymphoid aggregates in the nasal cavity and about the ostia⁽¹⁰⁾. When the primary site can be determined, it is often found to be the maxillary or ethmoid sinuses. Like carcinomas, lymphomas have a tendency to invade surrounding structures. The pattern of spread and bone destruction is largely dependent on the site of origin within the sinus. Lesions in the anterolateral infrastructure tend to invade the lateral inferior wall or grow through dental sockets. Lesions of the medial infrastructure readily invade the thin medial wall into the nasal cavity. Posterior infrastructure lesions erode into the infratemporal fossa. Extension of lesions into the orbit occur either directly through the roof of the maxillary sinus, indirectly via the ethmoids and lamina papyracea or by way of the infratemporal fossa and then through the infraorbital fissure⁽¹³⁾.

As with their epithelial counterparts, the prognosis of midfacial lymphomas depends greatly on the histological features and stage at the time of diagnosis⁽¹⁴⁾. Staging is difficult, and three staging systems are commonly employed (Ann Arbor, TNM-AJCC and the Wollner)⁽¹⁵⁾. The prognosis is worse in adults and in patients with lymphadenopathy, larger primary tumours, tumours of T-cell origin and dissemination during treatment⁽¹⁶⁾. Death occurs in approximately 50% of patients with local relapse and systemic involvement, often in other extranodal sites⁽¹⁷⁾.

The most effective treatment is surgical resection. Early lesions may be cured by surgery alone, but for most postoperative radiotherapy is required. Extension of the cancer to the skull base, nasopharynx or ethmoidal sphenoid sinus contraindicates surgical excision⁽¹³⁾. Other options include combined modality therapy (chemotherapy and radiotherapy).

This case report highlights the importance in considering neoplasia as a cause of mucocoele arising in paranasal sinuses, and also the importance of CT scanning in diagnosis.

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