The Journal of Laryngology and Otology
March 1990, Vol. **104**, pp. 217–221

Visual dysfunction: Abnormal sign of benign spheno-ethmoidal lesions

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Abstract
Isolated visual dysfunction with or without proptosis is rarely described as a sign of benign sphenoethmoidal lesions. Five patients with benign swelling of the sino-orbital region and related visual defects are reported. Cases include one patient with fibrous dysplasia, two patients with mucocoeles and two patients suffering from aspergillosis of the sinuses. A review of the literature and discussion of the cases is presented.

Introduction

Orbital cellulitis due to direct spread of infection from contiguous paranasal sinusitis is a well recognized syndrome. Visual dysfunction may occur with or following signs of marked proptosis, oedema of the eyelid and chemosis of the conjunctiva. On the other hand orbital apex syndrome secondary to adjacent sphenoethmoidal sinusitis with marked visual loss and ophthalmoplegia but with minimal proptosis or signs of anterior segment inflammation is rarely described (Kjoer, 1945).

We report five cases of benign lesions involving the posterior group of paranasal sinuses associated with reversible visual field defects with or without deterioration of their visual acuity. In only one patient did vision and proptosis not improve following surgical treatment. Whereas isolated visual dysfunction with or without proptosis as the initial sign of benign swelling arising in the sphenoethmoidal region was poorly documented in the ENT literature, more reports were collected from the ophthalmic literature. This report highlights the importance of visual examination and visual field acuity in otherwise innocent lesions in the sphenoethmoidal region.

Case report

Case 1

Blurring of vision in the right eye was the cardinal symptom of this 18-year-old male. He had suffered from right sided nasal obstruction for the previous four years and an abnormal protrusion of the right eye ball for one year. Examination revealed a contracted visual field with normal visual acuity. A hard swelling covered by intact mucosa was found filling the right nasal cavity and extending into the choana.

A CT scan showed very dense ground glass bone formation filling the entire nasal cavity. The medial orbital wall was displaced laterally and reduced to almost one third of its normal orbital circumference. The mass also involved the right ethmoid, sphenoid and maxillary sinuses and completely obliterated the frontal sinus (Fig. 1). The patient's visual field returned to normal one month after surgical excision of the tumour, which proved to be fibrous dysplasia.

Case 2

A 46-year-old male complained of disturbance of vision for the previous four months. Several ophthalmologic consultations were indecisive until later temporal contraction of the right visual field was noted. His visual acuity was 6/6. A CT scan of the head revealed encroachment of the optic nerve by a rounded circumscribed swelling 3 × 3 cm which occupied the posterior ethmoid air cells (Fig. 2). Excision of the swelling, which proved to be a mucocoele, and decompression of the optic nerve was performed. One month later the visual field returned to normal (Fig. 3).

Case 3

This 27-year-old female was admitted with a two month history of blurring and gradual diminution of vision together with abnormal protrusion of the right eye ball. She had undergone a left Caldwell-Luc operation six years previously. A swelling with an egg-shell crackling sensation, was felt in the floor of the right frontal sinus and was found to displace the eyeball downwards and laterally. The nose was filled with multiple soft polyps. The right visual acuity was 5/60 and the left 6/9. The right visual field showed concentric contraction. A CT scan showed a mass involving the paranasal sinuses causing extensive destruction of the bony compartments and the right medial orbital wall. There was also tumour extension intracranially involving both frontal lobes of

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Accepted for publication: 20 December 1989.

https://doi.org/10.1017/S0022215100112320 Published online by Cambridge University Press

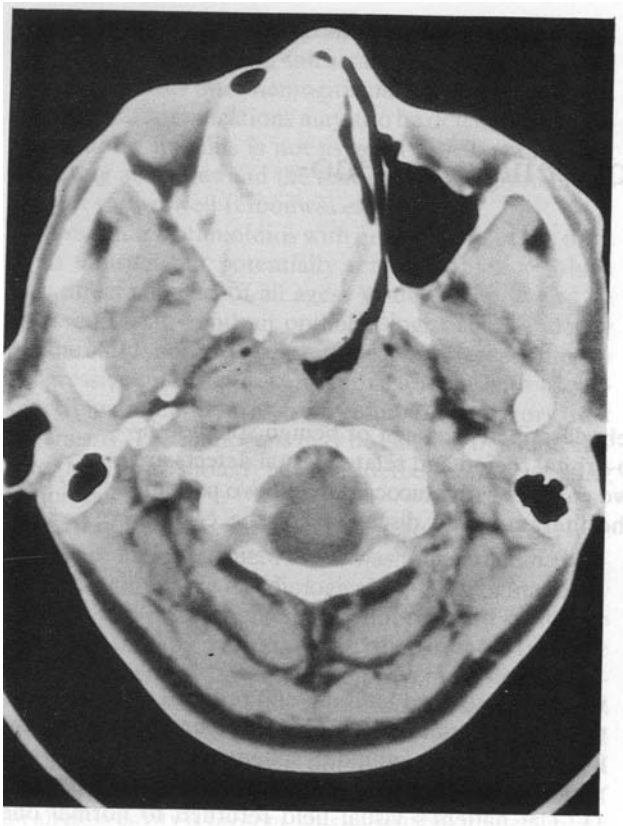


FIG. 1
CT scan showing extensive fibrous dysplasia displacing the medial orbital wall.

the brain. Magnetic resonance imaging confirmed the previous findings and a diagnosis of polypoid rhinosinusitis associated with a mucocoele with massive bilateral intracanial extension (Fig. 4) was made.

The patient underwent a bilateral fronto-ethmoido-sphenoidectomy. A massive mucocoele was found involving the frontal, ethmoids and sphenoid sinuses on both sides. The sinus mucosa was markedly oedematous and the cavity was filled with thick tenacious material. The posterior wall of both frontal sinuses was dehiscent. Histopathological examination and culture studies excluded fungal infection and also confirmed the radiographic diagnosis.

The patient has been attending regular follow-up examination for the past year. The visual acuity has considerably improved (6/9 in the right eye), the visual field returned to normal, and the nose is free of any recurrent polyps.

Case 4

A 40-year-old female complaining of long-standing nasal obstruction presented with left proptosis. She had undergone nasal polypectomy five times, previously and an intranasal ethmoidectomy. The nose was found to be filled with bilateral multiple polyps more so on the left side. Ophthalmological examination revealed a visual acuity of 5/60 in the left eye and 6/9 in the right with a limitation of the left peripheral field which had not been noticed by the patient.

A CT scan revealed a soft tissue density mass filling the left maxillary, sphenoid, frontal and ethmoid sinuses with encroachment on the orbital cavity causing lateral displacement of the medial rectus muscle (Fig. 5).

The patient underwent a left fronto-spheno-ethmoidectomy and a Caldwell-Luc procedure. Thick brownish mucoid discharge was trapped within the sinuses which were lined by markedly oedematous, polypoid mucosa. Aspergillus fungus was isolated from the excised contents. The patient was given a course of Amphotericin B and was put on systemic and local steroids for the next six months. The visual field was normal on follow-up examination one month after surgery.

Case 5

A 28-year-old female had undergone an external ethmoidectomy elsewhere one year previously. This was complicated in the early post-operative period by the development of a frontal pyocoele and later by right fistula formation. Two weeks post-operatively she underwent revision surgery with closure of the fistula. The patient was diagnosed as suffering from fungal sinusitis and was put on steroid therapy. Unfortunately the patient then complained of considerable diminution of vision.

Later the patient complained of a right sided proptosis, nasal obstruction, intermittent epistaxis and frontal headache. Irregular soft polypoid masses surrounded by brownish thick tenacious secretion were found to be filling the right nasal cavity. The patient could only perceive light in the right eye whilst the field of vision could not be traced.

A CT scan revealed a widely destructive mass involving the frontal, ethmoid and sphenoid sinuses with extension to the right orbital cavity.

Right external fronto-spheno-ethmoidectomy was performed with almost complete eradication of the disease. Tissue studies confirmed the diagnosis of Aspergillosis which was treated with intravenous Amphotericin B and steroids. No recurrence in the nose

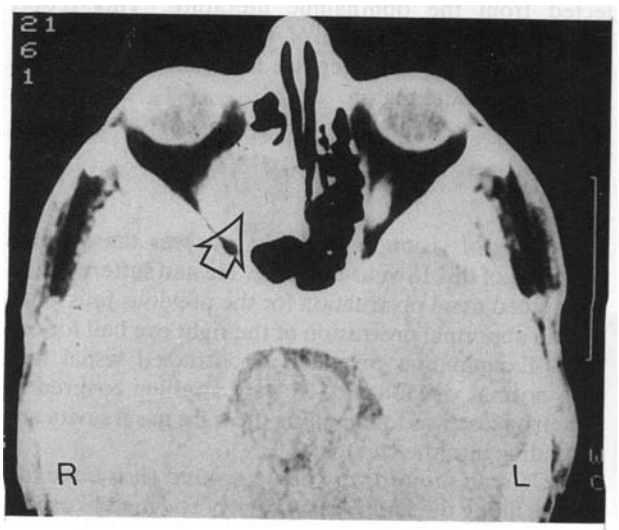


FIG. 2
CT scan showing mucocoele of the right posterior ethmoid sinuses (Case 1).

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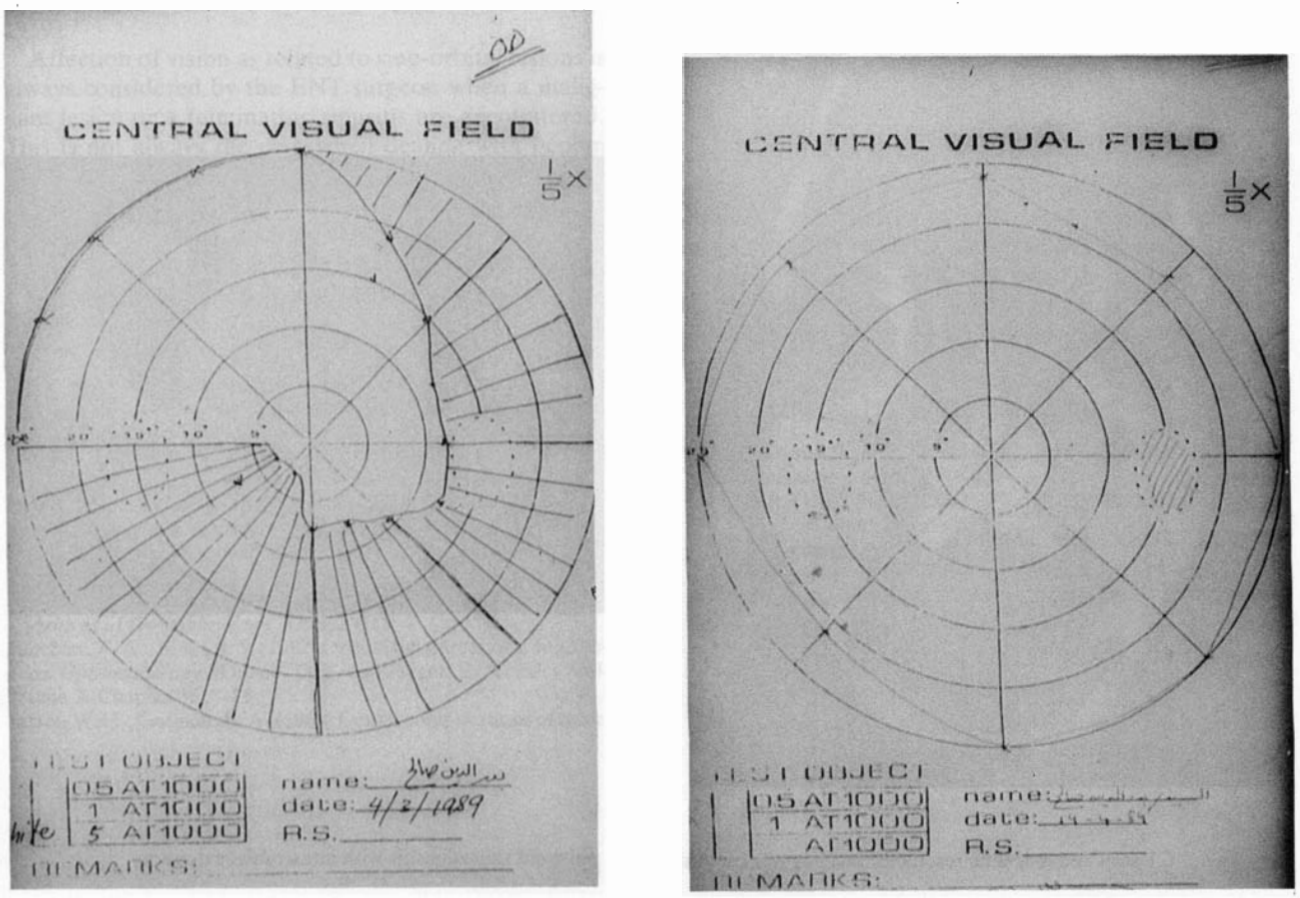


FIG. 3
Visual field of the same patient (Case 1) before and one month after surgery.

has been reported in the first two post-operative months but the proptosis and vision have not improved.

Discussion

Visual dysfunction secondary to adjacent posterior ethmoidal or sphenoidal sinusitis rarely occurs without associated orbital signs. With the exception of a malignant neoplasm, orbital cellulitis concomittant with paranasal sinusitis (mostly sphenoethmoiditis) is the commonest cause of sinus related visual dysfunction. There are many reports in the literature describing orbital apex syndrome due to sinus infection (Kronschnabel, 1974).

Jarret and Gutman (1969) described a patient with right paranasal sinusitis and acute unilateral visual loss which improved completely on medical treatment. Slavin and Glaser (1987) reported three cases of symptomatic sphenoethmoidal sinusitis in which severe irreversible visual loss overshadowed or preceeded signs of inflammatory orbital disease. They defined this condition as posterior orbital cellulitis (POC) syndrome.

This report describes five patients with POC due to different benign paranasal sinus pathologies. With the exception of one patient they all developed unilateral visual dysfunction which improved following surgical treatment. Pathology included fibro-osseous dysplasia, localized posterior ethmoid mucocoele, polypoidal rhinosinusitis with mucocoele and aspergillosis of the sinuses.

Fibro-osseous lesions cover a group of conditions where normal bone is replaced by collagen and varying amounts of osteoid tissue. Such masses may vary from extensive disfiguring tumours to small virtually unnoticed localized swellings. Involvement of the orbit with proptosis is unusual but might indicate early surgical intervention (Ramsey *et al.*, 1968).

Fu and Perzin (1974) reported a series of 64 fibro-osseous lesions of the paranasal sinuses. Modes of presentation were headache, facial asymmetry and painless swelling of the cheek and orbital regions. Moore (1969) has reviewed the ophthalmic literature on eight patients, including two of his own, presenting with ophthalmic symptoms. Proptosis developed fairly rapidly over a few weeks to months. In addition to exophthalmos, downward displacement of the globe as well as optic atrophy also occurred in the case of sphenoid lesions. Sassin and Rosenberg (1968) studied fifty patients with craniofacial disease. Visual loss from compression of the optic nerve was seen in eight out of the ten patients with radiographically demonstrable optic canal involvement.

Mucocoeles of the paranasal sinuses are benign, expansile lesions. They expand gradually, eventually eroding the bony walls of the sinus. Occasionally mucocoeles may have a widely destructive nature and may extend intracranially, as in case 3 (Close and O'Connor

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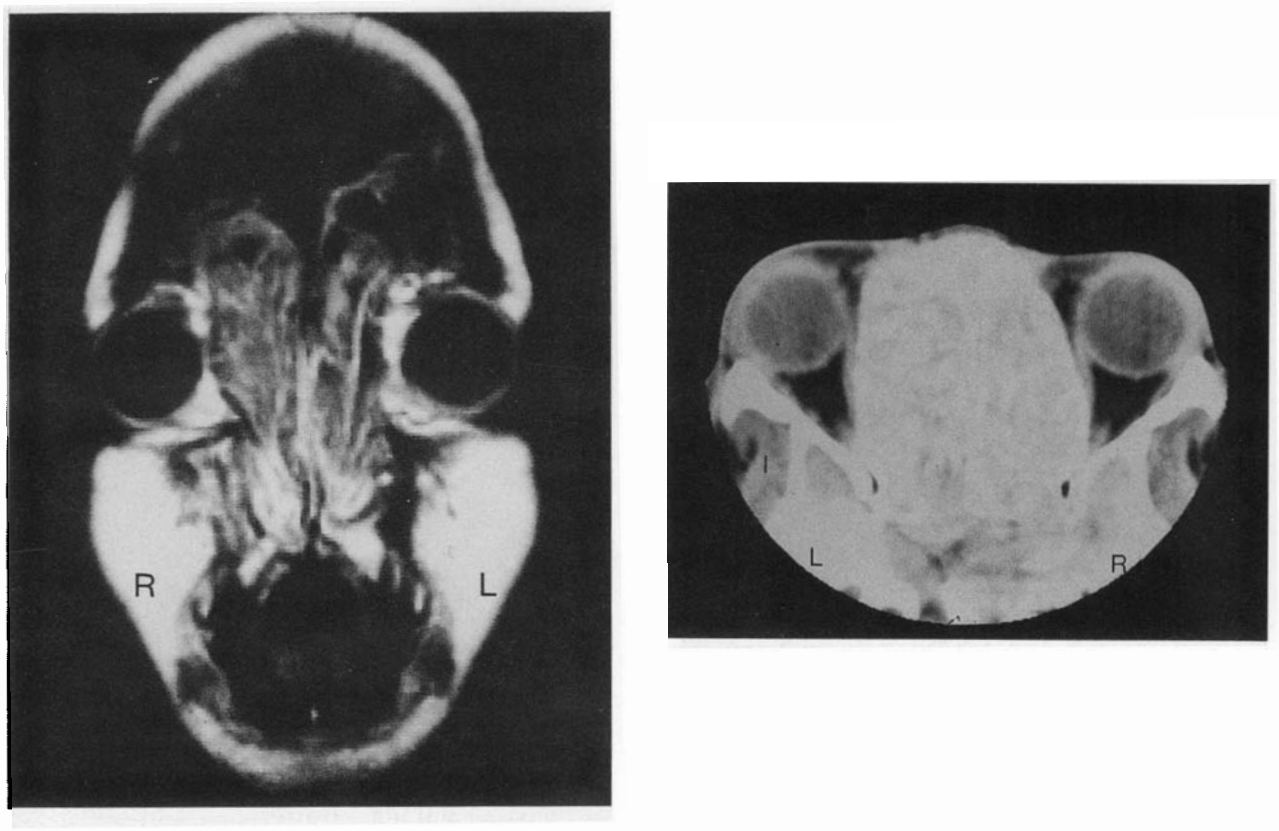


FIG. 4
CT scan and magnetic resonance imaging of Case 4 showing polypoid rhinosinusitis with intracranial extension.

1983; Schaeffer *et al.*, 1988). Very commonly, following a path of least resistance, the mucocoele projects into the orbit characteristically producing slowly progressive painless unilateral proptosis. Whereas the fronto-ethmoidal muocoele commonly produces proptosis and rarely involves the visual pathway, the opposite is true of the sphenoid mucocoele (Jakobiec and Jones, 1982).

Because of the way sphenoidal muocoeles expand, at times it cannot be established whether a sphenoidal mucoele developed within the sphenoid and spreads to the posterior ethmoids or vice-versa. Thus it may be preferable to term them sphenoethmoidal mucocoeles. Such mucocoeles may expand concentrically in all directions or may preferentially displace the intracavernous carotid artery, spread superiorly to elevate the planum sphenoidale, flatten the groove of the sella turcica, displace the pituitary, elevate and compress either optic nerve or chiasma and destroy the optic canals (Valvassori and Putterman, 1973). Despite a great deal of distortion of the optic nerve and canal, very good visual acuity may still be retained (case 2). Optic atrophy and visual disturbances are extremely rare with frontal and ethmoidal mucocoeles (Jakobiec and Jones, 1982).

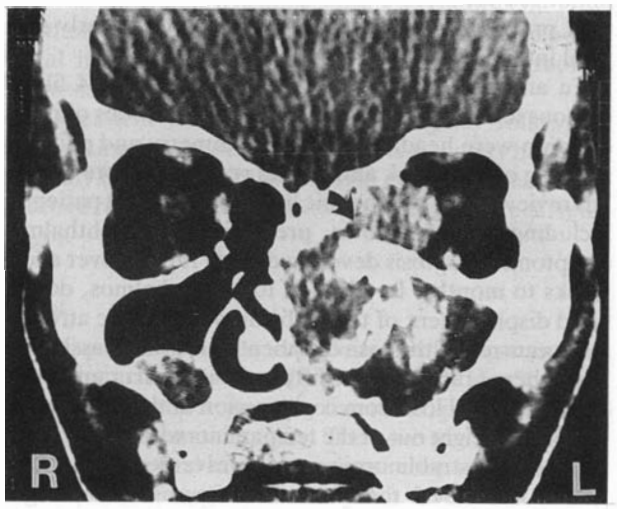


FIG. 5
CT scan showing displacement of the medial rectus muscle and the left orbital cavity by a mass almost filling the left group of paranasal sinuses.

Once thought of as a rare disease, aspergillosis of the nose and sinuses is being more frequently recognized. As a paranasal sinus disease, aspergillosis presents in several forms: non invasive, invasive fulminant and allergic aspergillus sinusitis (Sarti *et al.*, 1988).

The aspergillus organism is opportunistic, resides in the oropharynx and the sinuses, and is innocent most of the time. The aspergillus infection develops in a sinus and spreads to the orbit where it is usually a granulomatous fibrosing disease. It can, though, be a non-granulomatous fibrosing disease with abscess formation in patients who cannot mount a delayed hypersensitivity response.

Proptosis develops in those patients who have the granulomatous form. An orbital abscess associated with sinus disease may not produce proptosis and the patient may feel well. Once the sinus infection has elicited a fibrotic response in the orbit, the optic nerve is damaged with nerve head swelling and engorgement of the central retinal vein. In the granulomatous fibrosing form, as in case 5, there is a non-reducible proptosis because of the

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unyielding fibrous tissue behind the globe (Green *et al.*, 1969).

Affection of vision as related to sino-orbital lesions is always considered by the ENT surgeon when a malignant lesion or a fulminating sinusitis are encountered. This is not always the case when dealing with benign swellings in this region. Visual examination should be performed for all patients with lesions in the sphenoethmoidal area. This will provide valuable information as to the extent of the disease in the orbit and will be a better assessment both surgically and medicolegally.

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