CASE REPORT

Compressive optic neuropathy in the contralateral eye secondary to ethmoid sinus carcinoma

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Abstract

A 42-year-old gentleman presented with painless proptosis and diplopia in right eye. Vision and fundus were normal in both eyes. CT scan of orbits and brain revealed advanced ethmoid carcinoma on the right side. MRI of brain and orbits showed the tumour encroaching into right orbit, involvement of medial rectus and extension into right frontal lobe. Surgical excision of the tumour through bicoronal craniotomy by neurosurgeon, endoscopic-guided craniofacial resection by ENT surgeon and partial exenteration of right eye by ophthalmologist were done. Post operative radiotherapy was given. After 4 months tumour recurred causing rapid loss of vision in the left (contralateral) eye due to compressive optic neuropathy. Visual recovery was good following surgery and intravenous dexamethasone. Chemotherapy was given. However, patient died at home following another recurrence of the tumour.

Keywords: Optic neuropathy; Ethmoid carcinoma; Proptosis.

INTRODUCTION

Squamous cell carcinoma of the ethmoid sinus is a rare form of tumour and the symptoms are indistinguishable from that of inflammatory disease. Tumours of the nasal and paranasal sinuses constitute 0.2% to 0.8% of systemic malignancies and the maxillary sinus is the most common affected site. Although ethmoid sinus carcinoma is rare, it accounts for approximately 30% of paranasal sinus carcinoma, and squamous cell carcinoma is the most prevalent histopathologic type. The high mortality rate and poor prognosis are related to the late diagnosis which is caused by the early symptomatic latency of these tumours. Compressive optic neuropathy caused by sinusoidal undifferentiated carcinoma of ethmoid sinus was reported by Chen et al. The tumour was involving the cribriform plate, ethmoid sinus bilaterally, upper nasal cavity with intracranial extension into the frontal lobe. There was compressive optic neuropathy in their patient. Kains et al reported a 57-year-old man who presented with bilateral compressive optic neuropathy due to orbital compression by an ethmoid sinus adenocarcinoma. They stated that eye symptoms are as a result of the mass effect of the tumour leading to displacement of the
globe (non axial proptosis) or infiltration of the orbital structures. Tumour mass may penetrate the periorbita and infiltration of extraocular muscles may lead to restriction of ocular movements causing diplopia. Direct infiltration onto the optic nerve will cause diminution of vision, while involvement of sensory nerves gives rise to pain or reduced sensation in the infra orbital region. We report a case of young gentleman who presented to the eye clinic with proptosis in one eye due to ethmoid carcinoma on that side. Following the surgical treatment and post operative radiotherapy, there was a recurrence of the tumour causing compressive optic neuropathy in the other eye (contra-lateral side).

CASE REPORT
A 42-year-old gentleman presented to the eye clinic on 6/7/2006 with a history of painless and progressive protrusion of the right eye of one month duration with associated double vision on the left gaze. He had difficulty in reading. He also complained of right nasal discharge. There was no history of fever, loss of weight or appetite or any constitutional symptoms. He is taking treatment for hypertension since 2 months. There is no other significant medical illness.

The visual acuity of both eyes was 6/6. The right eye was proptosed in the inferolateral direction measuring 22 mm on the Hertel’s exophthalmometer when compared to 16 mm reading of left eye (Figure 1). The extraocular movement of the right eye was restricted superiorly and medially with diplopia in the said positions. The optic nerve function (visual acuity, papillary reaction to light, colour vision) of both the eyes was normal. The slit-lamp examination of the anterior segment of both eyes was normal. However, the intraocular pressure of the right eye by Goldmann tonometry was 15 mmHg in the primary position and 21 mmHg in the upgaze and that of the left eye was 15 mmHg and 19 mmHg respectively. The fundus examination showed of pink optic discs with well defined margin, cup-disc ratio of 0.3 and normal macula in both eyes.

Computed tomography of the brain and orbits revealed a large soft tissue mass arising from the ethmoid extending into the frontal lobe and retro-orbitally on the right side. Magnetic resonance imaging (MRI) showed the tumour encroaching into the right orbit (Figure 2), involvement of the right medial rectus (Figure 3) and extension into the right frontal lobe (Figure 4).
He was started on gutt timolol 0.5% bd for the right eye to lower the intraocular pressure. He was promptly referred to the neurosurgical and otorhinolaryngology team for joint management. The intraocular pressure was reduced to 18 mm Hg in right eye. The patient underwent a bicoronal craniotomy, excision of the tumour and calvaria reconstruction of the orbit. Intraoperatively, the anterior cranial fossa was explored by the neurosurgical team and it was found that the right orbit had been eroded by the tumour with invasion into the adjacent right frontal dural layer and the inferior part of the right frontal lobe. The tumour was excised and sent for histopathological examination, which confirmed the diagnosis of poorly differentiated squamous cell carcinoma. The otorhinolaryngology team performed an endoscopic-guided craniofacial resection of the ethmoidal tumour which was found to have arisen from the entire ethmoidal sinuses extending into the base of the skull. The ophthalmology team carried out partial exenteration (sparing the eyelids for cosmetic purpose) of the right eye to facilitate the endoscopic resection of the tumour. Post operatively, the surgical wound healed well and the patient was discharged from the hospital one week after the surgery. The vision, anterior segment and fundus were normal in the left eye. The patient was given radiotherapy for six weeks (external beam radiation, 54 Gy in 30 fractions delivered by a linear accelerator). Four months after completion of radiotherapy, he was admitted for rapid loss of vision in left eye of one week duration. The vision in the left eye was 6/60 and pupil was sluggishly reacting to light. Colour desaturation test showed grossly affected optic nerve function. Fundus examination showed pink disc with clear margin, no retinal haemorrhages or cotton wool spots. Macula was normal. CT orbits and brain showed the recurrence of the tumour invading the apex of left orbit (Figure 5).

He underwent an endoscopic trans-sphenoidal resection of the tumour 4 days later. Intraoperatively, the tumour was found extending posteriorly into the sphenoidal sinus. The patient was given I.V. dexamethasone 4 mg tds for 1 week. He progressed well and regained his vision to 6/12 on the fifth postoperative day. Chemotherapy with cisplatin and 5-flurouracil was administered. Two months later, telephonic call to his home revealed that he suffered another recurrence of the tumour and succumbed to the disease in September 2007.
DISCUSSION
Squamous cell carcinoma of ethmoid sinus may invade the orbit in preformed pathways such as infraorbital fissure and ethmoidal foramina. It may also invade the orbit through the neurovascular structures such as the ethmoidal, infraorbital and optic nerve, or by direct invasion. In this patient, the orbit was invaded by direct extension destroying the bone. The most common clinical presentation of sinonasal tumours are similar to those of benign and inflammatory sinus diseases and this delays the patients from seeking early treatment. The ethmoid sinus carcinoma has a tendency to spread insidiously to an advanced stage before causing symptoms and signs. Despite the extensive local growth, these tumours have a low propensity for lymphatic or haematogenous dissemination. The patients may initially experience nasal obstruction, rhinorrhoea, facial pain and epistaxis. Invasion of the orbit by the tumour tissue displaces the eyeball anterolaterally causing proptosis, as seen in this patient. Imaging is important for the assessment in patients with suspected orbital invasion. Plain sinus radiographs shows erosion of the bony orbit in 80% of the cases. Computed tomography scanning provides a better imaging of the bony framework but does not demonstrate the invasion of the soft tissue of the orbit. Magnetic resonance imaging provides an accurate evaluation of the actual soft tissue invasion. However, soft tissue invasion must be confirmed during surgery by the frozen section analysis.

The aims of the treatment are the oncologic cure, functional rehabilitation, cosmesis and the preservation of quality of life. The advanced stage at the time of diagnosis justifies aggressive surgical treatment with craniofacial approaches. Generally surgery alone offers a low control rate because resection with wide margins is rarely feasible due to tumour extent and the proximity of critical structures. Alvarez et al reported that radiotherapy alone gives poor prognosis for recovery (21% survival after 5 years). Therefore, definitive surgical resection and postoperative radiotherapy are recommended in the management of sinonasal malignancies. Jiang et al reported that 35% (12 out of 34) of patients with ethmoid sinus carcinoma had symptoms related to orbital involvement (diplopia, orbital pain, impaired vision, proptosis or periorbital swelling, inner canthus mass, watering of eye). They observed that presence of histologically proven dura mater invasion was associated with a high local recurrence rate. In our patient, the invasion of the dura mater was found at the first surgery itself and thus, the prognosis was anticipated to be very poor for long term survival.

Even with aggressive primary treatment, recurrence was common in squamous cell carcinoma of the ethmoid. Lavertu et al reported a 48% recurrence rate with an average time to recurrence of 9.9 months. Myers et al found that local recurrence was the most common, followed by distant, and then regional metastasis. Despite advances in imaging and the treatment, there continues to be a high mortality rate with carcinoma of the paranasal sinuses.

Diminution of vision or loss of vision in patients of ethmoid carcinoma could be due to radiation optic neuropathy or compressive optic neuropathy. In a study of 48 cases of malignant tumours of the nasal cavity and ethmoid and sphenoid sinus, Parsons et al reported 16 (33.3%) developed unilateral blindness secondary to radiation retinopathy or optic neuropathy which can be anticipated because of irradiation to the ipsilateral eye. Four patients in their study (8.3%) developed bilateral blindness after a long time (17, 35, 46, and 90 months) following treatment owing to optic neuropathy. The nature of radiation optic neuropathy is usually devastating. Most affected eyes have a final vision <6/60 and many are completely blind. Spontaneous improvement never or almost never occurs in patients with typical radiation optic neuropathy.

Absence of optic disc edema, retinal haemorrhages and cotton wool spots; and improvement of vision in the contralateral eye following surgery for local recurrence of the tumour support the diagnosis of compressive optic neuropathy in our patient.
The loss of vision in the contralateral eye was due to compression of the optic nerve because of spread of the tumour into the apex of the left orbit following its recurrence after surgery and radiotherapy. Ethmoid sinus carcinomas have a tendency for extensive local invasion but a low propensity for lymphatic and haematogenous spread. Hence, local recurrence is the main cause of cancer-related death\textsuperscript{11}. The main cause of death in our patient also was local recurrence despite of postoperative radiotherapy and chemotherapy.

References