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CASE REPORT

Sinonasal adenocarcinoma with aggressive ocular symptom

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ABSTRACT

BACKGROUND. The malignant sinonasal tumour is very rare. Sinonasal adenocarcinoma comprises only 10-20% of all primary malignant sinonasal tumours. The commonest type is the maxillary squamous cell carcinoma. It commonly presents with nasal blockage, nasal discharge and epistaxis during the early stage. Headache and blurry vision may occur at an advanced stage when it has invaded the brain, the eye or the optic nerve.

CASE REPORT. We present a 63-year-old patient with acute progressive worsening unilateral blurry vision and headache for 1 month. Epistaxis with anosmia developed only later. The patient had a neuroimaging by both Computed Tomography (CT) scan and Magnetic Resonance Imaging (MRI) scan which showed a skull base tumour, but early biopsy was inconclusive. He underwent combined transcranial and transsphenoidal tumor debulking in view of clinical impression of olfactory neuroblastoma. The histopathological examination showed adenocarcinoma. He was sent for postoperative radiotherapy.

CONCLUSION. We highlighted that a patient with sinonasal adenocarcinoma may present initially with the symptom of invasion to neighbouring structures prior to the local symptom.

KEYWORDS: sinonasal tumour, adenocarcinoma, ocular symptom, intracranial spread.

INTRODUCTION

Malignant sinonasal tumours are very rare, comprising only 3% of the head and neck tumours. Sinonasal adenocarcinoma arises from the respiratory epithelium and the seromucinous glands at the sinonasal tract. Common presenting symptoms include nasal blockage, rhinorrhea and epistaxis. It may invade adjacent structures such as the brain, the eye and the optic nerve, leading to disease progression complication such as blindness and seizures, necessitating a radical surgery.

Typically, the sinonasal tumour carries a poor prognosis especially in case of squamous cell carcinoma, adenocarcinoma and undifferentiated carcinoma.

CASE REPORT

A 63-year-old Indian man presented with rapid progressive visual changes in his right eye over a one-month period, associated with headache and giddiness. His acute presentation was due to epistaxis and anosmia 1 week prior. No symptoms of nasal blockage, seizures or weakness were noted.

He had underlying hypertension and diabetes. He used to smoke. He never worked as a woodworker.

On examination, he was alert, normotensive (122/78mmHg). Pupils were equal and reactive to light at 3mm bilaterally. Relative afferent pupillary defect (RAPD) was negative. Visual acuity was impaired significantly, only with the ability to count fingers on the right and 6/24 on the left. He had anosmia, with normal muscle strength and sensory perception. There was no palpable cervical lymph node.

He was initially diagnosed with bilateral, non-arteritic, anterior ischaemic optic neuropathy (NAION). Cerebral Computed Tomography (CT) scan showed a skull base tumor arising from the planum sphenoidale with suprasellar, intracranial and extraconal extension (Figure 1).

The intranasal tissue biopsy was inconclusive, showing a necrotic exudate and abundant neutrophils.

Magnetic resonance imaging (MRI) of the brain showed a mass measuring 5.3x6.8x6.3cm, which appeared hypointense on T2W and isointense on T1W, with the epicentre in the superior nasal cavity and the ethmoid sinuses (Figure 2). Superiorly, it extended into the anterior cranial fossa towards the planum sphenoidale, with marked perilesional oedema and dural en-

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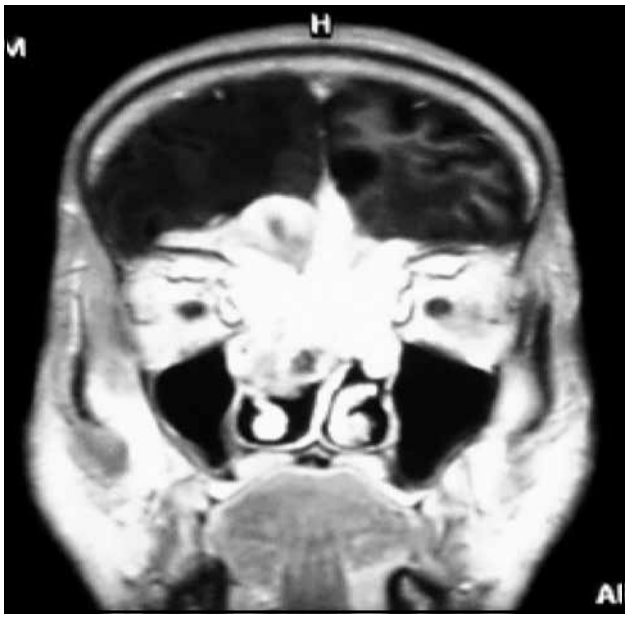

Figure 1 Coronal CT scan showed a large ill-defined heterogeneously enhancing expansile lesion extending to the frontal lobes.

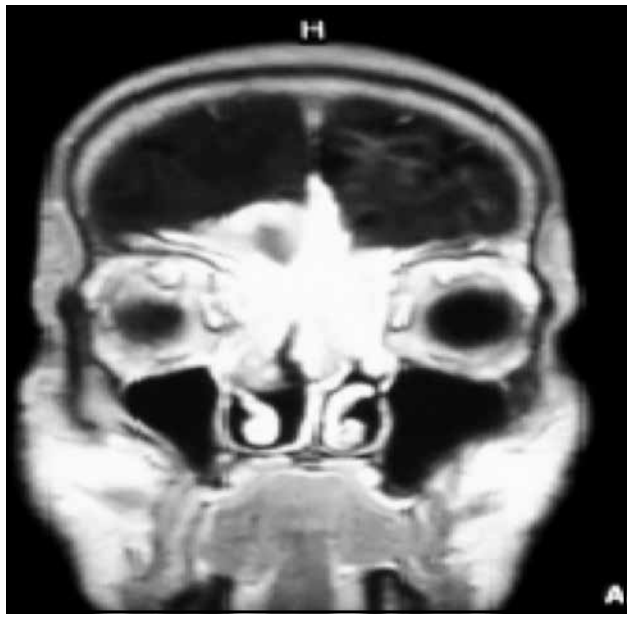

Figure 2 The mass appeared to be isointense at the right frontal lobe level on T1W MRI, extending intracranially.

hancement. The tumour engulfed the distal part of both intracranial optic nerves, intracanalicular optic canals and the right orbital apex. Extraocular muscles were spared, except the right superior rectus muscle. The inferior extension was in the nasal cavity. There was no hydrocephalus or midline shift. The common carotid and the internal carotid arteries were patent.

A diagnosis of Stage IVb (T4b N0 M0, AJCC 2017) sinonasal tumour was made.

He was advised for combined transcranial transsphenoidal debulking of the tumor, but he refused. He presented again 2 weeks later with bilateral eye blindness, behavioural changes as well as executive function and mental impairment. RAPD was positive and the fundoscopy revealed optic atrophy of the right fundus and papilledema of the left fundus.

He underwent combined transcranial, transnasal endoscopic approach tumor debulking which necessitated bifrontal craniectomy, septostomy, medial orbitectomy and sphenoidectomy. Intraoperatively, the tumor appeared moderately vascular, yellowish with extensive erosion of the frontal bone, dura, the cribriform plate, the sphenoid sinus and the frontal lobe. It adhered to the frontal sinus and superior sagittal sinus.

Post tumor resection, the brain remained swollen, necessitating bone flap removal and placement of an external ventricular drainage (EVD), for CSF drainage post operatively.

The histological examination showed moderately differentiated adenocarcinoma. There were fibrocollagenous tissues infiltrated by malignant cells forming a glandular pattern and papillae. These malignant cells were moderately pleomorphic, with vesicular nucleus and some having prominent nucleoli.

His postsurgical CT scan of the brain did not show residual bleed, with only minimal perilesional oedema (Figure 3).

His recovery was complicated with development of VAP, which was managed expectantly. He was discharged well on day 19 postoperatively with GCS score of E3M6V4.

He defaulted his oncology appointment for radiotherapy.

DISCUSSIONS

Malignant sinonasal tumours are rare, comprising only 3% of all head and neck malignancies[1]. Only 10-20% of primary malignant sinonasal diseases are sinonasal adenocarcinomas[2]. They predominantly affect men (male to female: 6.8 to 1), with the average age of occurrence at 65.7 years[3].

The complexity of the sinonasal tract anatomy which lies near the skull base makes a potential for tumoral invasion to the orbit, the optic nerve and the brain. Common presenting symptoms are nasal blockage (71.9%), epistaxis (53.1%) and rhinorrhea (15.6%). These symptoms mimic rhinosinusitis. In this case, there was a red herring, masking the more sinister sign of tumor extension. Less common symptoms are orbital signs (9.4%), pain (9.4%), skin damage (9.4%) and neurological signs (6.2%)[3].

A study by Johnson L et al. showed that 64% of patients presented with an orbital symptom at the initial presentation; they were mostly in the maxillary squamous cell carcinoma group. Common presenting complaints in these groups were facial or eye pain, facial swelling, nasal congestion and epistaxis[4]. Our reported case is unique as his presenting complaints were orbital and intracranial symptoms. His nasal symptom developed later.

Distant metastasis is rare with only 4.3%[5]. Sinonasal adenocarcinoma has a reported 5-year survival rate of 45%, whereas squamous cell carcinoma, undifferentiated carcinoma, melanoma and adenoid cystic carcinoma have a survival rate of 29%, 31%, 53%, 100%, respectively[6].

Non-arteritic anterior ischaemic optic neuropathy (NAION)

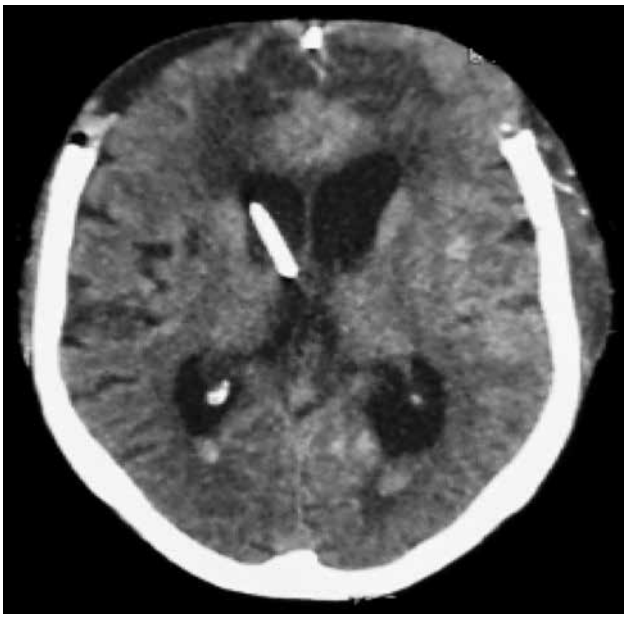

Figure 3 The mass appeared to be isointense at the right frontal lobe level on T1W MRI, extending intracranially.

is the commonest cause of acute optic neuropathy due to acute ischemia of the optic nerve in people of above 50 years old[7]. Typically, NAION presents with sudden reduction in vision, with relative afferent pupillary defect and optic disc oedema[7], much like this case, that eventually exhibits all the characteristics and may contribute to the array of masking symptoms, making an accurate and early diagnosis a challenge. It carries poor visual prognosis and the contralateral eye may be affected in up to 15% of the cases[7]. We propose a neuroimaging for all patients above 50 years old with acute optic neuropathy to ensure skull base tumors are ruled out.

The treatment modalities for sinonasal adenocarcinoma are surgery, radiotherapy, chemotherapy or in combination.

Challenges in the surgical management for malignant sinonasal diseases are partly due to their close proximity to critical structures (brain, optic nerve and eye). The aim of surgery is for maximum tumour resection while minimizing complications. Surgical approach options include endoscopic the transnasal approach, transcranial approach or a combination of both. The open method includes transcranial, lateral rhinotomy, midfacial degloving, Caldwell-Luc and craniofacial resection, which can be performed as a single procedure or in combination. The choice of surgery depends on the primary tumor location and tumoral extension. Surgery in comparison to chemo- or radiotherapy may cause complications such as wound infection, injury to the globe, the optic nerve and the extraocular muscle, cerebrospinal fluid (CSF) leaks and pneumocephalus. A pooled analysis performed by Meccariello G et al. showed a lower rate of complications in the endoscopic technique. However, this data has selection bias, as smaller tumours are treated by endoscopic approach[8].

Patients treated only with radiotherapy are due to unresectable locally advanced tumour. Adjuvant radiotherapy after partial surgery showed 85% local control rate[9].

Chemotherapy can be used as neoadjuvant for cytoreduction before surgery. A study done by Brasnu et al. on cases of a locally advanced tumour reaching or invading the skull base showed local control in 65.7% of the cases[10].

Our patient presented with locally advanced skull base malignancy. The aim of surgery in this case was cytoreduction, tissue diagnosis and radiosensitization.

Postoperative deterioration in his conscious level may be due to postoperative oedema, bleed or pre-existing frontal lobe invasion. Other contributing factors are electrolyte imbalance as well as progression of the disease. However, his postsurgical CT scan and blood parameters were stable.

CONCLUSIONS

Recognition of orbital and intracranial symptoms for sinonasal as well as other skull base tumours is crucial for rapid appropriate management. Delay in diagnosis may sacrifice vital structures leading to functional and life-threatening complications. Hence, an urgent CT scan is mandatory in a patient presenting with orbital and intracranial symptoms.

Conflict of interest: The authors have no conflict of interest.

Contribution of authors: All authors have equally contributed to this work.

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