Acute proptosis with optic neuropathy secondary to primary sinonasal neuroendocrine carcinoma – a case report and review of the literature

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Introduction

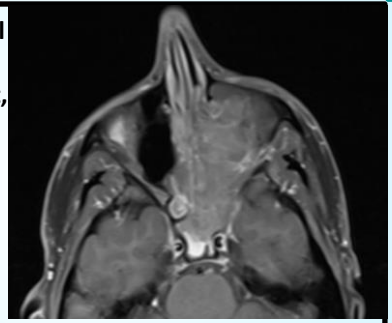
Primary sinonasal neuroendocrine carcinoma is a rare and aggressive form of neuroendocrine tumour with high recurrence rate and tendency to metastasise. We present a rare case of advanced sinonasal neuroendocrine tumour (NET) with acute ophthalmic presentation from a district general hospital in London, UK

Case Presentation

A 36-year-old male presented with 2 weeks left proptosis and blurred vision on a background of 4 months left nasal discharge and cheek pain. Examination confirmed left sided 2mm proptosis, RAPD, ishihara 11/17, visual acuity (VA) of 0.1 Logmar (vs. 0.0 right), and globally reduced extraocular movements (EOM).

Case Investigations

Figure 1: MRI of a large left nasal cavity mass extending into the left maxillary sinus and left orbit, compressing on the left optic nerve, and extending intracranially.



Nasal biopsy:T4b poorly differentiated nasal carcinoma with neuroendocrine differentiation.

Case management

After 1 cycle of induction chemotherapy with cisplatin and etoposide, all ocular signs resolved. Disease remission was achieved with further chemoradiotherapy. The patient is stable at 4 years.

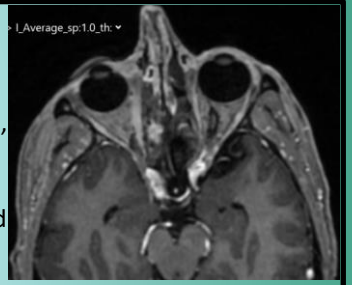


Figure 2: Most recent MRI showing no disease

Discussion

Primary sinonasal NET is rare. There are 6 reported cases with primary orbital involvement (Agrawal et al. 2023). Our case is unique in the complete resolution of ocular signs with chemotherapy. Contrary to the literature, which suggests that combined treatment with surgery is associated with better survival, our patient showed excellent response to combination chemotherapy and radiotherapy alone. Whereas these tumours frequently recur locally and metastasise, our patient has been disease-free for 4 years.

Learning points

Acute presentation with orbital signs requires history to include sinonasal symptoms, a multidisciplinary approach, and timely imaging and biopsy where appropriate.