Orbital paraganglioma: case report and review of the literature

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Abstract
Paragangliomas of the orbit are extremely rare. A case of an orbital paraganglioma, including the first magnetic resonance imaging description of this tumour is described here. The patient underwent surgery with gross total removal of the tumour and relief of his initial chief complaint of visual blurring. The differential diagnosis and therapeutic options for the management of this tumour are discussed.

Paragangliomas are neoplastic growths of the non-chromaffin paraganglionic tissue which may arise in the abdomen, thorax, or head and neck region. Though paragangliomas of the head and neck are not uncommon they are rarely found in the orbit, where the site of origin is thought to be the ciliary nerve. Indeed only 27 cases have been reported in the literature. The present report gives details of such a case and provides the first compilation of all previously described orbital paragangliomas.

Case history
The patient is a 55-year-old man who presented to the Medical Center Hospital of Vermont with intermittent visual blurring during the preceding 6 months. Physical examination revealed slight hypertropia and exotropia of the right eye. Vision was 20/25 OS and 20/20 OD. Pupils were equal and reactive. Spontaneous venous pulsations were absent on the right and normal venous pulsations were present on the left.

The visual field on the right showed concentric constriction of isopters and a small superior defect at 40° out. Colour vision was noted as 10/14-12/14. A right afferent pupillary defect of about 1-2 log units was noted.

Computed tomography (CT) of the orbits revealed a homogeneous mass lesion of 2 cm separate from and superolateral to the optic nerve. Prominent orbital veins were identified (Fig 1).

The patient underwent a right lateral orbitotomy with visualisation of a firm, well circumscribed mass with an adherent vascular capsule. Frozen section examination suggested the diagnosis of a meningioma. Therefore the operation was terminated after partial internal debulking of the mass with a plan for postoperative radiation therapy.

Permanent microscopic sections of the tumour revealed nests of polygonal or spindle cells surrounded by a delicate fibrovascular stroma forming the so-called 'zellballen pattern' (Figs 2 and 3). The tumour cells were moderate in size and round to polygonal in shape. The nuclei were oval, slightly hyperchromatic, and pleomorphic. Mitoses were rare. Immunohistochemical staining revealed focal positivity of the
tumour cells for neuron specific enolase and chromogranin. Ultrastructural examination revealed occasional tumour cells containing scarce to many neurosecretory granules (Fig 4).

Six months postoperatively, a magnetic resonance (MR) scan (Fig 5) of the right orbit was obtained and demonstrated a 2-0 cm by 1-8 cm intracanal mass that displaced the optic nerve medially and inferiorly. The typical prominent vessels and 'salt-and-pepper' appearance of the MR image as described for paragangliomas of more typical locations were present. Because of residual tumour and a recurrence of his diplopia, the patient underwent a right fronto-orbital craniotomy as described by Jane et al with removal of the right orbital apex mass. Pathology was again consistent with the diagnosis of paraganglioma. The procedure was terminated when tumour-free margins were obtained.

Discussion
Paraganglioma of the orbit is a rare tumour with 27 such cases reported in the literature. The pathological differential diagnosis of orbital paraganglioma includes meningioma, metastatic adenocarcinoma (especially renal cell carcinoma) alveolar soft part sarcoma, alveolar rhabdomyosarcoma, and hemangiopericytoma. Ultrastructural demonstration of neurosecretory granules in the chief cells of paraganglioma is diagnostic.

The MR images allowed further characterisation of the tumour mass, in that the salt-and-pepper appearance of the matrix is strongly suggestive of the diagnosis of paraganglioma. Prominent vessels might occur in association with other intracanal orbital masses, especially cavernous haemangioma, although this has not yet been described in reported cases. Intraorbital meningiomas usually do not have grossly enlarged vessels that can be demonstrated by CT or MRI. Optic nerve glioma would not have a salt and pepper matrix or prominent vascularity. Intraorbital metastases are usually not sufficiently hypervascular to have grossly enlarged vessels associated with them.

While surgery is the treatment of choice for paragangliomas radiation therapy for unresectable or incompletely resectable tumours, unencapsulated tumours, and for recurrence has been generally recommended. Tumour regression has been demonstrated using this procedure though at least one case report failed to note any effect of postoperative radiotherapy (5000 cGy) on tumour size. However there was no discussion regarding the effect of radiation therapy on the patient's symptomatology.

In summary this report reviews the pertinent literature to date and presents the pathological and radiological appearance of an orbital paraganglioma including the first MR description to our knowledge. Although rare in this location a paraganglioma should be considered in the differential diagnosis of a vascular orbital mass.

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