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.¹ Indeed only 27 cases have been reported in the literature.¹⁻²³ 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.

Figure 1 Enhanced axial CT scan of the orbit. There is a 1.5 cm × 2.0 cm well defined homogeneously enhancing mass that lies above the optic nerve in the apex of the muscle cone.

Figure 2 Nests of epithelioid cells in 'zellballen' pattern, characteristic of paraganglioma (haematoxylin and eosin, ×400).

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.²⁴

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

Figure 3 Reticulin stain outlining nests of cells in paraganglioma (reticulin, ×200).

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Figure 4 Dense core granules (arrow) within the cytoplasm of tumour cells ($\times 11\,500$).

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 intraconal 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.²⁵ Because of residual tumour and a recurrence of his diplopia, the patient underwent a right fronto-orbital craniotomy as described by Jane *et al*²⁶ 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.¹⁻²³

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 characterisa-

tion of the tumour mass, in that the salt-and-pepper appearance of the matrix is strongly suggestive of the diagnosis of paraganglioma.²⁵ Prominent vessels might occur in association with other intraconal orbital masses, especially cavernous haemangioma, although this has not yet been described in reported cases.²⁷⁻²⁹ Intra-orbital 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.³⁰

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.³¹ Tumour regression has been demonstrated using this procedure¹⁰ though at least one case report failed to note any effect of postoperative radiotherapy (5000 cGy) on tumour size.² 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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- 1 Archer KF, Hurwitz JJ, Balogh JM, Fernandes BJ. Orbital nonchromaffin paraganglioma. A case report and review of the literature. *Ophthalmology* 1989; **96**: 1659-66.
- 2 Ahmed A, Dodge OG, Kirk RS. Chemodectoma of the orbit. *J Clin Pathol* 1969; **22**: 584-8.
- 3 Amemiya T, Kadpya M. Paraganglioma of the orbit. *J Cancer Res Clin Oncol* 1980; **96**: 169-79.
- 4 Benfu W. Orbital chemodectoma: clinical and pathologic analysis of two cases. *Clin Med J* 1981; **94**: 419-22.
- 5 Deutsch AR, Duckworth JK. Nonchromaffin paraganglioma of the orbit. *Am J Ophthalmol* 1969; **68**: 659-63.
- 6 Fisher ER, Hazard JB. Nonchromaffin paraganglioma of the orbit. *Cancer* 1952; **5**: 521-4.
- 7 Gonsalves CG, Briant TDR. Chemodectomas of the head and neck. *J Can Assoc Radiol* 1979; **30**: 109-15.
- 8 Hughes LW, Ambrose A. Retro-orbital adrenal rest tumor. *JAMA* 1944; **126**: 231-2.
- 9 Juhasz J, Nagy F, Selymes Z. Über das nicht-chromaffine paragangliom der orbita. *Ophthalmologica* 1962; **144**: 103-10.

Figure 5 A, B Axial and coronal MR scans of the orbits demonstrate a well defined tumour mass in the muscle cone, sitting above and lateral to the right optic nerve (A; TR 2500 ms, TE 30 ms). Punctate and serpiginous structures with flow voids represent prominent arteries and veins associated with the tumour (A). Note also the lateral orbitectomy defect in A and B. Coronal T1-weighted spin echo image (B; TR 600 ms, TE 20 ms) confirms intraconal location of the mass.

Fig 5A

Fig 5B

- 10 Lack EE, Cubilla AL, Woodruff JM, Farr HW. Paragangliomas of the head and neck. *Cancer* 1977; 39: 397-409.
- 11 Lattes R, McDonald JJ, Sproul E. Non-chromaffin paraganglioma of the carotid body and orbit. *Ann Surg* 1954; 139: 382-4.
- 12 Litricin O. Paragangliome non-chromaffine de l'orbite. *Ann Oculist (Paris)* 1970; 203: 585-92.
- 13 Mathur SP. Non-chromaffin paraganglioma of the orbit. *Int Surg* 1968; 50: 336-9.
- 14 Niraukari MS, Greer CH, Chaddah MR. Malignant non-chromaffin paraganglioma in the orbit. *Br J Ophthalmol* 1963; 47: 357-63.
- 15 Pautique L, Girard PF, Audibert J. Une tumeur orbitaire exceptionnelle: le paragangliome non-chromaffin. *Ann Oculist (Paris)* 1962; 195: 131-41.
- 16 Rippey JJ, Rohm GF, van den Heever CM, DeWit LJD. Paraganglioma of the orbit - an exercise in diagnosis. *S Afr Med J* 1981; 60: 148-50.
- 17 Tesinsky P, Kurka R. Maligni achromafinni paraganglion orbity. *Cesk Oftalmol* 1963; 19: 325-8.
- 18 Thacker WC, Duckworth JK. Chemodectoma of the orbit. *Cancer* 1969; 23: 1233-8.
- 19 Valach V. Die extraadrenalen paragangliome. *Zentralbl Allg Pathol* 1957/58; 97: 251-64.
- 20 Vara Thorbeck R, Morales Valention OI, Morales MR. Non-chromaffin paraganglioma of the orbit. *Zentbl Chir* 1986; 111: 46-9.
- 21 Venkataramana NK, Sastry Kolluri VR, Raj Kumar DU, Vasudev Rao T, Das BS. Paraganglioma of the orbit with extension to the middle cranial fossa: case report. *Neurosurgery* 1989; 24: 762-4.
- 22 Wu B. Orbital chemodectoma: clinical and pathologic analysis of two cases. *Chin Med J (Engl)* 1981; 94: 419-22.
- 23 Zhang L, Wang KL, Wu KL. Chemodectoma of the orbit: report of a case. *Acta Acad Med Wuhan* 1982; 2: 60-2.
- 24 Smith JL, Vuksanovic MM, Yates BM, Bienfang D. Radiation therapy for primary optic nerve meningiomas. *J Clin Neuro Ophthalmol* 1981; 1: 85-99.
- 25 Olsen WL, Dillow WP, Kelly W, Norman D, Brant-Zawadzki M, Newton TH. MR imaging of paragangliomas. *Am J Neuroradiol* 1987; 148: 201-4.
- 26 Jane JA, Park TS, Pobereskin LH, Winn HR, Butler AB. The supraorbital approach: a technical note. *Neurosurgery* 1982; 11: 537-42.
- 27 Bilaniuk LT, Atlas SW, Zimmerman RA. Magnetic resonance imaging of the orbit. *Radiol Clin North Am* 1987; 25: 509-28.
- 28 Fries PD, Char DH, Norman D. MR imaging of orbital cavernous hemangioma. *J Comput Assist Tomogr* 1987; 11: 418-21.
- 29 Mafee MF, Putterman A, Valvassori GE, Campos M, Capek V. Orbital space-occupying lesions: role of computed tomography and magnetic resonance imaging an analysis of 145 cases. *Radiol Clin North Am* 1987; 25: 529-59.
- 30 Peyster RG, Shapiro MD, Haik BG. Orbital metastasis: role of magnetic resonance imaging and computer tomography. *Radiol Clin North Am* 1987; 25: 647-62.
- 31 Sonneland RPL, Scheithauer BW, LeChago J, et al. Paraganglioma of the cauda equina region. *Cancer* 1986; 58: 1720-35.