University Hospital of Wales, Heath Park,
Cardiff CF14 4XW, UK
Tel: +44 2920 747 747;
Fax: +44 2920 748 300.
E-mail: hjbunting11@yahoo.com

Eye (2006) **20,** 1449–1450. doi:10.1038/sj.eye.6702315;
published online 3 March 2006

Sir,

Choroidal ganglioneuroma in a patient with neurofibromatosis type 1: a case report

Ganglioneuromas are rare, benign tumours, forming part of a spectrum of tumours arising from primordial neural crest cells in the sympathetic nervous system.[1] However, they can dedifferentiate into the malignant neuroblastoma and ganglioneuroblastoma.[1] Choroidal ganglioneuroma is extremely rare.[2] Herein, we report a patient with neurofibromatosis type 1 (NF-1) who underwent evisceration for a painful blind eye, subsequent histopathological examination (HPE) revealing a clinically unsuspected choroidal ganglioneuroma.

Case report

A 11-year-old boy presented with severe pain in his congenitally enlarged blind left eye. He had undergone a glaucoma filtering surgery at 1 year of age, details of which were unavailable.

Visual acuity in the normal right eye was 6/6. There was an upper eyelid plexiform neurofibroma on the left side and multiple facial *café-au-lait spots* (Figure 1, left). The left eye was buphthalmic with multiple iris Lisch nodules and a cataractous lens precluding fundus examination. Intraocular pressure was 34 mmHg. Computed tomography (CT) scan showed an enlarged left eye. The greater wing of the sphenoid was hypoplastic (Figure 1, right). A clinical diagnosis of NF-1 with a painful blind left eye was made. As the patient desired cosmetic improvement, an evisceration with silicone implant with subsequent prosthesis fitting was considered. At surgery, there was no evident abnormal intraocular tissue.

HPE revealed thickened choroid with a cellular lesion, consisting of bundles of spindle cells admixed with clusters of ganglion cells, with no atypia, necrosis, or pleomorphism (Figure 2, left). The ganglion cells showed abundant cytoplasm, a large vesicular nucleus with prominent nucleoli and stained brightly with neuron-specific enolase (Figure 2, centre). These features were diagnostic of a benign choroidal ganglioneuroma. Immunohistochemistry with neurofilament (Figure 2, right) confirmed the presence of ganglion cells and with S-100 confirmed the neural bundles.

Comment

Patients with NF-1 have inactivation of the 17q11 tumour suppressor gene predisposing to tumours of neural crest origin.[3] A decrease in neurofibrin production with subsequent increase in Ras-GTPase activity causes cellular proliferation and mitosis.[3]

Choridal ganglioneuroma in NF-1 is very rare, with MEDLINE search revealing only one published case report.[2] In this patient also, enucleation for a painful

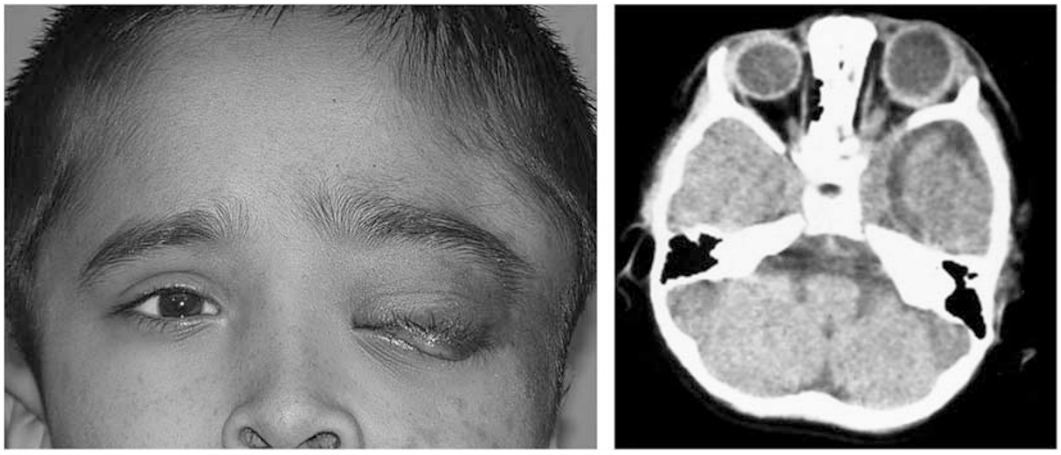

Figure 1 NF-1 with choroidal ganglioneuroma: external photograph of the face showing bupthalmos of the left eye and multiple *café-au-lait* spots on the face (left) and an axial CT scan showing enlargement of the left eye and absence of the greater wing of sphenoid (right).

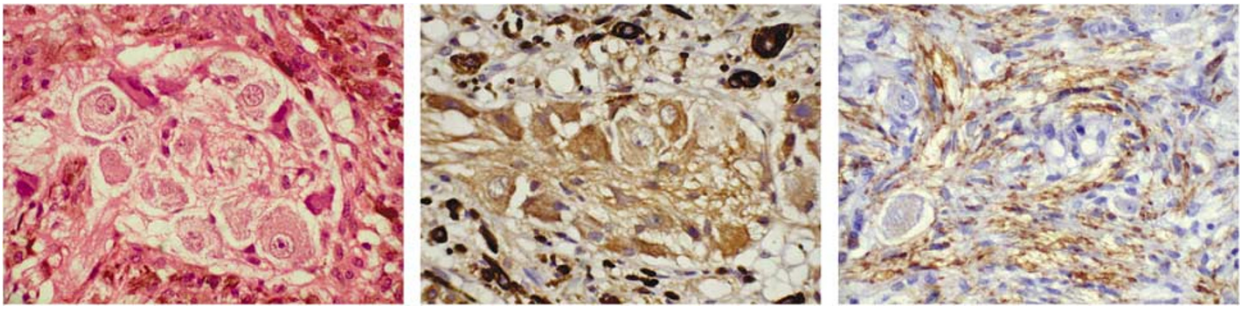

Figure 2 NF-1 with choroidal ganglioneuroma: histopathology of the choroidal mass showing a cellular lesion consisting of bundles of spindle cells admixed with clusters of ganglion cells (haematoxylin and eosin, ×500) (left), immunohistochemistry showing neuron-specific enolase staining of the ganglion cells (centre), and immunohistochemistry showing neural filament protein within the cytoplasm of the spindle cells (right).

blind eye revealed an unsuspected choroidal ganglioneuroma.

Ganglioneuromas are most commonly located in the posterior mediastinum (42%) and retroperitoneum (38%).[4] Reports exist of ganglioneuromas in various organ systems of the body in association with NF-1, with an associative relation being hypothesized.[1]

We report a patient with NF-1 who underwent evisceration with HPE demonstrating a hitherto unsuspected choroidal ganglioneuroma. Reports of local recurrences with malignant transformation are of concern, entailing close follow-up.[1]

References

1 Shimada H, Ambros IM, Dehner LP, Hata J, Joshi VV, Roald B *et al*. The International Neuroblastoma Pathology Classification (the Shimada system). *Cancer* 1999; **86**: 364–372.
2 Woog JJ, Albert DM, Craft J, Silberman N, Horns D. Choroidal ganglioneuroma in neurofibromatosis. *Graefes Arch Clin Exp Ophthalmol* 1983; **220**: 25–31.
3 Arun D, Gutmann DH. Recent advances in neurofibromatosis type 1. *Curr Opin Neurol* 2004; **17**: 101–105.
4 Hayes FA, Green AA, Rao BN. Clinical manifestations of ganglioneuroma. *Cancer* 1989; **63**: 1211–1214.

D Shome[1], GK Vemuganti[2] and SG Honavar[1]

[1]Department of Ophthalmic Plastic Surgery, Orbit and Ocular Oncology, LV Prasad Eye Institute, Hyderabad, India

[2]Department of Ophthalmic Pathology, LV Prasad Eye Institute, Hyderabad, India

Correspondence: SG Honavar,
Department of Ophthalmic Plastic Surgery,
Orbit and Ocular Oncology,
LV Prasad Eye Institute,
LV Prasad Marg,
Banjara Hills,
Hyderabad 500034, India
Tel: +91 40 30612622;
Fax: +91 40 23548271.
E-mail: honavar@lvpei.org

Financial support: Hyderabad Eye Research Foundation, Hyderabad, India

The authors have no financial interest in any of the methods or materials used in the study

Eye (2006) **20,** 1450–1451. doi:10.1038/sj.eye.6702320; published online 24 March 2006

Sir,

Intraoperative breakage of the mushroom manipulator tip during phacoemulsification

We report three cases of breakage of a mushroom manipulator tip at the end of phacoemulsification. The technique used was divide and conquer in all three cases.

Case reports

Case 1

At the end of the fourth segment removal, the mushroom manipulator tip was missing. Aspiration of soft lens matter was carried out, a posterior chamber lens was implanted, and the tip was detected floating inside the capsular bag beneath the implant. The capsular bag was filled with heavy ophthalmic viscoelastic device (OVD), the implant was tilted using a lens dialler, and the tip retrieved with vitreous forceps.