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Eye (2006) **20,** 1405–1406. doi:10.1038/sj.eye.6702259; published online 3 February 2006

Sir,

Solitary nasal neurofibroma presenting as compressive optic neuropathy

Neurogenic tumour of the nasal cavity is a rare entity and it is rarer still for it to present with visual problem. We like to report a rare case of solitary nasal neurofibroma, which came to light through optic nerve compression.

Case history

A 40-year-old Malay woman was referred by the primary care physician because of blurred vision in her left inferior visual fields for the past 2 months. Her only past medical history of note was hypertension, controlled with oral atenolol. At presentation, her vision was 6/6 in the right eye and 6/12 in the left eye but there was no afferent pupillary defect. The intraocular pressures in both eyes were not raised and there were no signs of glaucomatous discs. However, the left optic disc appeared pale. A visual field test was ordered which revealed a left central scotoma.

CT scan revealed a large solid midline nasal tumour (Figure 1). Superiorly the tumour eroded and left behind a thin superior wall of the sphenoid bone. However, the pituitary fossa and optic chiasm were intact. Laterally, it eroded through the ethmoid bones and sinuses. In the left orbit, the tumour can be seen to compress on the optic nerve. Inferiorly, the tumour extends into both nasal cavities and filled up the entire middle meatus. The nasal septum was destroyed. The source and nature of the tumour cannot be inferred from the scan. At the 6-week follow-up after the scan, her left vision had decreased to counting finger with an obvious relative afferent pupillary defect.

A nasal endoscopic biopsy of the lesion was performed by the ENT surgeon. The tumour appeared white with a firm consistency. Histologically, the specimen was made up of spindle wavy cells with basophilic nuclei (Figure 2). No malignant cells were seen. The initial diagnosis was that of a fibromatosis. The tumour was excised jointly by the neurosurgeon and the ENT surgeon via bifrontal craniotomy and transnasal approach,

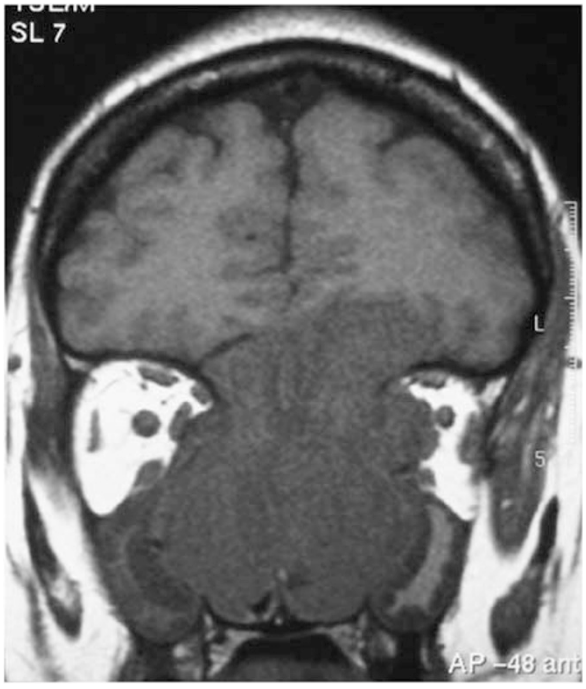

Figure 1 CT scan showing a large infiltrative midline lesion invading the left orbit and compressing the left optic nerve.

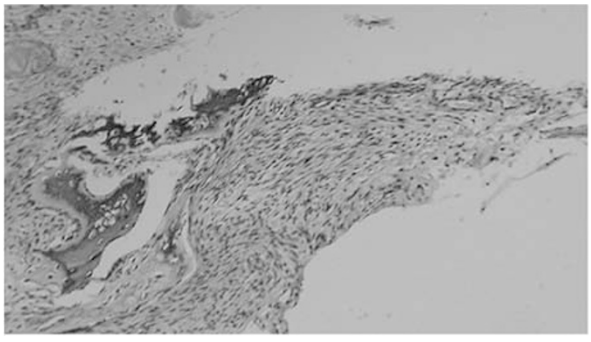

Figure 2 Biopsy specimen showing spindle wavy cells typical of neurofibromatosis (H&E staining).

respectively. During the procedure the wall of the sphenoid and ethmoidal and the nasal septum were removed as there were infiltrated and destroyed by the lesion. Immunohistochemical studies revealed that the cells of the tumour were immunoreactive vimentin, neuron-specific enolase and S-100 protein but negative for epithelial membrane antigen. From these and the clinical findings, the tumour was diagnosed as a neurofibroma.

Postoperatively, the patient made a good recovery and her vision improved to 6/12 with resolution of the relative afferent pupillary defect. Repeated serial CT scans showed the presence of residual tumour (Figure 3), which was confirmed on further nasal endoscopic biopsy.

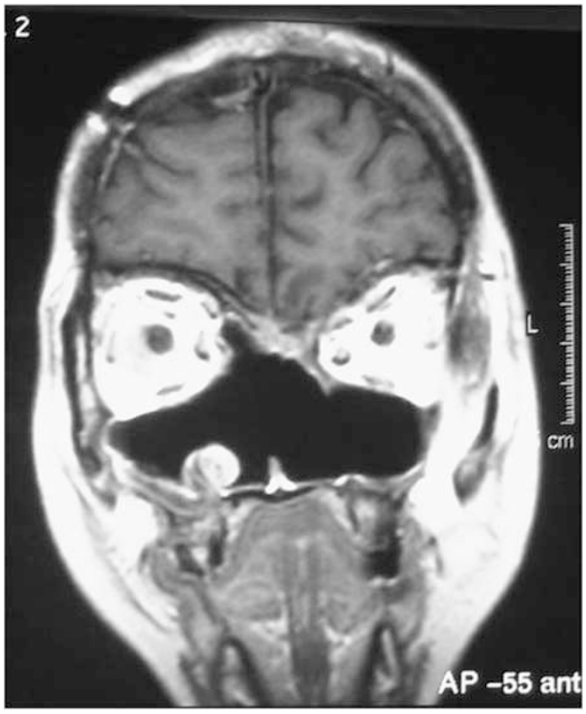

Figure 3 Postoperative CT scan at 6 months showing absence nasal septum and residual tumour.

However, there were no signs of growth at 18 months follow-up and the patient remained symptom free.

Discussion

Neurofibroma is a benign neurogenic tumour, which may occur singly or as a part of the syndrome of the neurofibromatosis (both neurofibromatosis type I and II). In the latter, the lesions are always multiple. Although 25–45% of neurofibroma occurs in the head and neck, only 4% are found within the nasal cavity.[1] Perzin *et al* revealed only six neurofibromas in a review of 4300 pathology specimen from the nasal cavity, paranasal sinuses, and nasopharynx.[2]

Neurofibroma in the nasal cavity often leads to nonspecific symptoms including nasal obstruction, cheek swelling, and epitaxis.[3,4] The tumour can also erode into the orbit[5] but to our knowledge there has not been any case reports in which the patient presents with compressive optic neuropathy as in this case. Radiologically, the tumour may resemble a malignant tumour because of the presence of bony destruction. The only reliable way to differentiate it from other tumours is through histological examination.[2]

Macroscopically the tumour appears as an encapsulated white mass with a firm consistency. Histologically, the tumour consists of wavy cells with basophilic nuclei with collagenous tissue. However, for an accurate diagnosis, the tumour needs to be subjected to immunohistochemical studies as the histological appearances tend to be nonspecific. Immunohistochemically, the cells of the tumour show immunoreactivity for vimentin, neurone specific enolase and S-100 protein.

The treatment for neurofibroma is surgical resection.[2] Extensive infiltration may make this difficult as is the case in our patient. However, even if incompletely excised recurrence usually takes many years. Regular follow-up is important as residual tumour may grow with time but malignant transformation is uncommon.

Our case shows that neurofibroma may present as an expanding midline nasal mass causing compressive optic neuropathy. This tumour should be considered in the differential diagnosis of any lesions arising from the nasal cavity, which secondarily invades the orbit.

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Eye (2006) **20,** 1406–1408. doi:10.1038/sj.eye.6702261;
published online 10 February 2006

Sir,

Pericentral pigmentary retinopathy: long-term follow-up

Pericentral pigmentary retinopathy is a rare disorder characterized by annular chorioretinal atrophy with varying degrees of bone spicule pigmentation extending temporally from the optic disk along the main retinal vessels in an arcuate fashion. Rarely, macular complications may be observed, such as bull'eye maculopathy and macular hole due to foveal ischemia and vitreomacular traction.[1] The prognosis varies, as the visual acuity and ocular findings appear stable in the autosomal recessive form[2] (OMIM[3] 268060) and progressive in the autosomal dominant form[4] (OMIM 180210).

Case report

A 14-year-old patient born to consanguineous parents was first seen in the Retina Unit of the University of Naples Federico II in 1987, complaining of night blindness. Patient's general health was good, metabolic and enzymatic profiles were within normal ranges, no relevant infectious disease was reported and history of exposure to toxic agents was negative. Visual acuity was 20/20 OU, while at fundus examination a bilateral peripapillary and pericentral chorioretinal atrophy with bone spicule pigmentation that spared the peripheral retina and the macula was noticeable. Fluorescein angiography showed bilaterally areas of pigmented epithelium atrophy along main retinal vessels, with medium to coarse pigment clumping. In both eyes automated perimetry showed an annular scotoma, the ERG was nearly extinct, Ishihara colour test was altered. Examinations of both parents were normal. During 18 years of follow-up the visual acuity of the patient has slowly worsened to 20/200 OD and 20/40 OS. At fundus examination progressive peripheral extension of chorioretinal atrophy and ring-shaped pigment, associated to a mild atrophy of the foveolar pigment epithelium have been observed. Periodic fluorescein angiography has shown wider peripheral atrophy of the retinal pigment epithelium and peripheral retinal nonperfusion. Multiple episodes of chorioretinal neovascularization arising in ischaemic areas have occurred. Thus, repeated argon laser photocoagulation of both ischaemic areas and neovessels has been performed (Figures 1 and 2). The electroretinographic responses have become unrecordable and automated perimetry has shown a progressive enlargement of the annular scotoma.

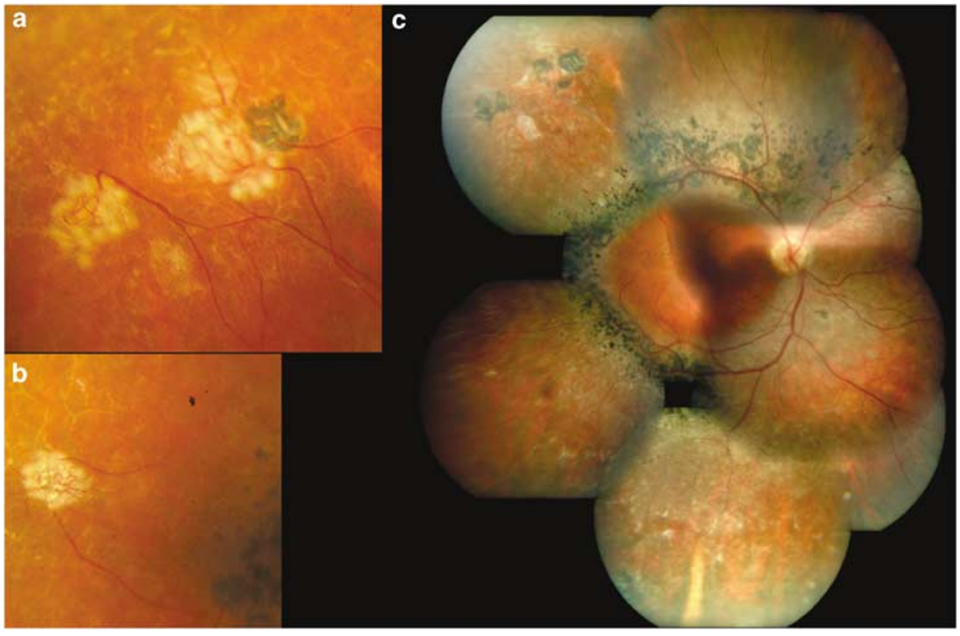

Figure 1 Pericentral pigmentary retinopathy. (a and b) Laser-treated chorioretinal neovessels of the right eye. (c) The colour map of the same eye shows the scars of photocoagulation in the supero-temporal periphery.