Optic nerve glioma and neovascular glaucoma: report of a case

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SUMMARY Neovascular glaucoma in a blind eye was a complication of an enlarging intraconal optic glioma in an 11-year-old girl. Venous stasis retinopathy was observed several months before the appearance of iris neovascularisation and elevated intraocular pressure. We suggest that venous stasis retinopathy in a patient with optic glioma is a reasonable indication for resection of the intraconal portion of the tumour.

Neovascular glioma is an unusual complication of optic nerve glioma. This report describes a young patient with monocular blindness from an enlarging intraconal optic glioma. After a progressive increase in proptosis she developed venous stasis retinopathy and neovascular glaucoma.

Case report

The patient, an 11-year-old girl, was first seen by her ophthalmologist in December 1978 after a school nurse discovered that she had defective vision in her left eye. Visual acuity in this eye improved from 6/7.5 +4 to 6/6 +4 with a −0.75 dioptre spherical lens. Vision in her right eye was 6/6 +4 unaided, but she had a 1+ afferent pupillary defect and mild swelling of the optic disc in this eye. The left disc was normal. Colour vision, extraocular movements, and exophthalmometer readings were normal. Examination with a slit-lamp showed no abnormalities. The intraocular pressure was 15 mmHg bilaterally. A visual field examination revealed a relative defect in the inferior nasal quadrant of the right eye. The left visual field was full.

A computerised tomography (CT) scan showed fusiform, intraconal enlargement of the right optic nerve to 2-3 times its normal diameter, the enlarged segment extending from immediately behind the globe to the orbital apex. The right optic canal was enlarged. There was no evidence of intracranial extension. The intracranial optic nerve, suprasellar cistern, and ventricles were normal. Tomograms showed enlargement of the right optic canal at both its orbital and intracranial ends; its orbital diameter was 1.5-2.0 mm greater than that of the left canal. The walls of the canal were normal. Pneumoencephalography with polytomography confirmed that the intracranial segment of the right optic nerve was normal in diameter. A glioma was suspected. A general physical examination revealed no signs of neurofibromatosis.

Three months later the patient complained of painless dimming of vision in the right eye. Visual acuity was 6/9 unaided, improving to 6/7.5 with correction; 1.5 mm of proptosis and a 4+ afferent pupillary defect were noted. Swelling and congestion of the right optic disc were noticeably greater than before. Visual field loss was more marked, with an inferior altitudinal defect to small targets.

In November 1980 visual acuity in the right eye had deteriorated to the extent that she could see to count fingers at 6 inches (15 cm) in one quadrant only. Right proptosis had increased to 3 mm. Intraocular pressure was 17 mmHg bilaterally. Swelling of the right disc had increased, and there was marked venous stasis retinopathy, with optociliary shunt vessels and intra-retinal haemorrhages around the optic nerve head. The left disc was normal (Fig. 1).

A month later she had 2 episodes of acute pain in her right eye, each of which resolved spontaneously. She was admitted to hospital for further examination. She was unable to perceive light and had 12 mm of axial, irreducible proptosis in her right eye. The eye showed marked ciliary injection. The right pupil was fixed in response to light stimulation and was widely dilated...
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Fig. 1 Fundus photographs of both eyes. The right disc is swollen and shows marked venous stasis retinopathy, optociliary shunt vessels, and intraretinal haemorrhages around the optic nerve head. The left disc is normal.

Fig. 2 Right eye showing marked ciliary injection and optic nerve glioma. It has been reported in 2 cases of small gliomas, following central retinal vein occlusion in one case and following anterior segment ischaemia in the other. The initial manifestation of an optic glioma in our patient was visual loss and swelling of the optic disc. Progressive enlargement of the tumour, observed clinically and by CT, was associated with (1) increasing proptosis, (2) decreased vision, (3) progressive optic atrophy and optociliary neovascularisation of the iris (arrows).

(10 mm). Examination with a slit-lamp revealed corneal oedema, a shallow anterior chamber, and extensive neovascularisation and atrophy of the iris (Fig. 2). The angle of the anterior chamber was completely closed by neovascular membrane over 360 degrees. Intraocular pressure 49 mmHg. The right disc was pale and swollen and showed signs of marked arteriolar narrowing, marked venous dilatation, scattered venous haemorrhages, and optic disc atrophy. A second CT scan showed enlargement of the intracanal optic nerve mass and contrast enhancement of the periphery of the tumour (Fig. 3). The suprasellar cistern and intracranial optic nerves were normal.

Through a Kronlein orbitotomy the anterior portion of the tumour was excised from directly behind the globe and as far posteriorly as the exposure would allow. Histopathological examination showed anaplastic astrocytoma; the central retinal vein was not occluded, and the central retinal artery was not identified. Postoperatively the intraocular pressure was 42 mmHg. The retinal arterioles were bloodless and barely visible; the retinal veins contained blood but were small. The patient's eye remains pain-free with topical medical therapy.

Discussion

Neovascular glaucoma is a rare complication of an optic nerve glioma. It has been reported in 2 cases of small gliomas, following central retinal vein occlusion in one case and following anterior segment ischaemia in the other. The initial manifestation of an optic glioma in our patient was visual loss and swelling of the optic disc. Progressive enlargement of the tumour, observed clinically and by CT, was associated with (1) increasing proptosis, (2) decreased vision, (3) progressive optic atrophy and optociliary neovascularisation of the iris (arrows).
We believe that iris neovascularisation and neovascular glaucoma in our patient were the result of the adverse effects of the enlarging tumour on the ciliary and retinal circulation and subsequent retinal ischaemia. Venous stasis retinopathy, marked retinal arteriolar narrowing, and extensive atrophy of the iris, together with a markedly dilated pupil, provided further evidence of retinal and ciliary ischaemia.

Preservation of the eye is a major objective in the management of orbital optic glioma.¹² Our case illustrates how this goal was compromised by the development of neovascular glaucoma while the patient awaited a cosmetic operation to relieve proptosis. Although this complication is very rare, we now believe that venous stasis retinopathy in a patient with an optic glioma is a reasonable indication for resection of the intraconal portion of the tumour.

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