CASE REPORT

Central Retinal Vein Occlusion in a Childhood Optic Nerve Tumour

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ABSTRACT

The authors describe the first paediatric patient with an aggressive optic nerve tumour of uncertain histology causing a central retinal vein occlusion, retinochoroidal collaterals, arteriovenous anastomoses, and peripheral retinal non-perfusion. He first presented with pale optic nerve head, followed by development of optic disc oedema a year later. Certain optic nerve tumours can present with central retinal vein occlusion in the paediatric age group.

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Introduction

The association of optic nerve tumour with optic disk oedema and retinal vascular occlusion has been reported infrequently. Tumours of the optic nerve are particularly uncommon in the paediatric age group and include predominantly gliomas and, rarely, optic nerve sheath meningiomas.¹ We describe a boy without known neurofibromatosis type 1 or 2 who had an optic nerve tumour that eventually caused unilateral blindness with a central retinal vein occlusion and retinochoroidal collaterals.

Case report

This boy was seen in the neuro-ophthalmology clinic at the age of 9 years with the report that he became aware of very poor vision in his right eye approximately 2 months before when he happened to cover his left eye. He had a history of substantial head trauma 3 years before, and the family brought with them a report of an ophthalmologic examination performed elsewhere at that time that described right eye trauma. The parents had noted that he developed an exotropia shortly after that trauma, although the child himself was not aware of diplopia or visual loss at that time. His

examination revealed no light perception on the right, with a 4+ relative afferent pupillary defect (APD) and moderate optic atrophy on that side. Afferent visual functioning and fundusoscopic examination was normal on the left. He had full ocular motility, with a modest comitant sensory exotropia on the right. A magnetic resonance imaging (MRI) scan showed mild diffuse thickening of the intracanal portion of the right optic nerve with bright signal on T2-weighted images (Figure 1A, B).

He returned 1 year later with the report of continued no light perception in the right eye, although his pupil on the right was minimally reactive to light and on occasion he squinted to a bright light in that eye. On dilated fundusoscopic examination, the right optic disc was mildly elevated and quite pale with an irregular outline, obvious venous congestion on the surface of the disc, blot haemorrhages along the superior and inferior arcades consistent with a central retinal vein occlusion (CRVO), and a few peripapillary arteriovenous anastomoses (Figure 2). Intraocular pressures measured 16.5 mm Hg on the right and 16 mm Hg on the left. He again had excellent afferent visual functioning on the left, with a normal fundus on that side. He was not proptotic on the right, although he had

Figure 1. Magnetic resonance imaging of the orbits and brain. (A and B) Initial magnetic resonance imaging (MRI) in 2013 at the age of 9. Axial and coronal sections of T2-weighted MRI of the orbits showed right optic nerve thickened, with high signal intensity (small white arrow) as compared with the normal contralateral side. Contrast study was not performed at that time. (C and D) Follow-up MRI study at the age of 10. Axial T1- and T2-weighted images showed significant thickening along the intraorbital portion of the right optic nerve, with flattening of the posterior globe around the optic nerve head. (E and F) Axial fat-suppressed T1-weighted MR images of the orbit shows the tram-track sign where the optic nerve (black dashed arrow) appears as a negative defect with a tram-track appearance in relation to the surrounding enhancement of the optic nerve sheath on either side (white dashed arrow). (G and H) Coronal images at the orbital apex showing that the optic nerve is relatively spared.

mildly increased resistance to retropulsion on that side. Extraocular motility was again full bilaterally, with a modest comitant sensory exotropia. A search for café au lait spots was unrevealing, and the remainder of his neurologic examination was unremarkable.

Fundus fluorescein angiography on the right showed slightly delayed laminar venous filling, staining of retinal veins, retinochoroidal collaterals, and peripheral non-perfusion on wide-field angiography (Figure 3). Optical coherence tomography demonstrated significant thickening of the

Figure 2. Colour fundus photography of the right eye. (Left panel) Colour fundus photo of the right eye demonstrated optic disk oedema, engorged and tortuous retinal blood vessels, and dot haemorrhages in all quadrants. (Right panel) Detailed image of the same eye showing retinochoroidal collaterals (arrow) and arteriovenous anastomosis (arrowhead).

Figure 3. Fundus fluorescein angiography of the right eye. (Left panel) Wide-angle fundus fluorescein angiogram of the right eye demonstrated staining of retinal veins in all quadrants and capillary non-perfusion in the retinal periphery. (Right panel) Posterior pole fluorescein angiogram of the same eye showed staining of the retinal vein (arrow), retinochoroidal collaterals at the temporal margin of the optic disk (arrowhead), and fluorescein leakage at the optic disk (thin arrow).

nerve fibre layer on the right compared with the left. B-scan ultrasonography documented an enlarged and elevated right optic nerve head with the “sheathing sign” but no obvious ocular or orbital mass.

Repeat MR imaging at this time showed diffuse, uniform enlargement of the intraconal portion of the right optic nerve, with flattening of the posterior globe in the region of the optic disc, similar signal intensity to the brain grey matter on T1-weighted image (WI) and T2WI, moderate homogeneous enhancement predominantly in the peripheral portion of the optic nerve and its sheath, and sharp demarcation, with the central, darker, non-enhancing optic nerve giving the appearance of “tram track” sign (Figure 1C–H). The canalicular and intracranial portions of the right optic nerve appeared normal. The left optic nerve, the optic chiasm, the retrochiasmal optic tracts, other orbital structures, and the brain all had a normal MRI appearance. There was a lack of the eighth cranial nerve involvement on MRI. These observations

were suggestive of either a right optic nerve glioma or optic nerve sheath meningioma. The patient’s prognosis and the benefits and risks of various investigative and treatment options were discussed with the child’s father; however, he refused additional studies or intervention.

Discussion

This boy followed an unusual clinical course. By report, he developed an exotropia without diplopia at approximately the age of 7 years, presumably because of reduced vision in the right eye of which he was unaware. When examined at age 9 years, the right eye was behaviourally blind, with a flat, moderately atrophic optic nerve head and minimal thickening of the optic nerve on MRI scan (Figure 1A, B). By 1 year later, he had a CRVO with right optic disc swelling in association with retinochoroidal collaterals, arteriovenous anastomoses at the margin of the disc and along the inferior arcade, and peripheral vascular non-perfusion (Figure 2). The right optic

nerve had increased considerably in size (Figure 1C–H), and probably the tumour located there caused constriction of the central retinal vein. Nevertheless, he had no proptosis, no restriction of ocular motility, no café au lait spots, and no history of additional general medical problems when last examined. Although the diagnosis of the most common optic nerve tumours can be made with neuroimaging, the diagnosis is not made until the orbit is explored and a biopsy of the nerve is obtained in some patients. The evolution of the MRI appearance in this patient was ambiguous, with characteristics that might imply either an optic nerve glioma (ONG) or an optic nerve sheath meningioma (ONSM). Unfortunately, the patient's father refused additional evaluation that might have been diagnostic.

Histologically, ONGs arise from glial cells within the optic nerve and remain closely associated with the optic nerve regardless of the pattern of growth so that there is involvement of both sheath and nerve substance without distinction between the two. Kinking of the tumour and cystic spaces within its substance are more common than a tubular pattern.¹ Additionally, most gliomas enhance wholly or in part after intravenous injection of a gadolinium-DTPA (diethylenetriamine pentaacetate), and it is thought that enhancing lesions are more metabolically active than those that do not enhance.² Gliomas of the anterior visual system in children are usually low grade, whereas malignant gliomas almost always occur in adults.^{3,4} Most ONGs are stable or slowly progressive^{5,6}; however, ONGs may demonstrate aggressive local expansion on occasion, leading to progressive deterioration in vision. Only two patients have been reported previously with an ONG causing venous stasis retinopathy, opticociliary shunt vessels, and/or rubiosis iridis with neovascular glaucoma.⁷ One patient presented with a CRVO associated with a biopsy-proven high-grade ONG; this patient underwent both radio- and chemotherapy, with resultant optic atrophy and no light perception vision.⁸ A second patient had an anaplastic optochiasmatic glioma and presented with central retinal vein and artery occlusions.⁹ Both patients were adults with malignant ONGs, and to the best of our knowledge, no paediatric patient has been reported with this retinovascular presentation. Routine early

detection and intervention for progressive ONG may prevent or delay significant pressure on the retinal vascular system, making a CRVO uncommon in current practice.

ONSMs are due to a proliferation of meningotheelial cells within the orbital or intracanalicular portion of the optic nerve sheath. They occur most commonly in adults (mean age at presentation of 40.8 years¹⁰), where they tend to be slow growing. Only 4% of ONSM patients are less than 20 years old, and as of 2012 only 14 patients had been reported with ONSMs under the age of 10, and none of these individuals had a CRVO.¹¹ These childhood tumours tend to be more aggressive, causing visual loss¹² and proptosis,¹⁰ even though the histology of these tumours does not appear more malignant than those found in adults.¹³ Approximately 30% of paediatric patients have had disk oedema, and a few have been documented with retinochoroidal collaterals,¹⁰ but none have been reported with a CRVO. Neuroimaging frequently exhibits the tram-track sign caused by circumferential calcification or enhancement adjacent to the optic nerve that is thought to be related to the linear spread of meningeal cells once the tumour has extended from its original location in the subdural space.¹⁴ Tram-track sign is due to a dense and thickened optic nerve sheath caused by the linear spread of tumour and is believed to be a sign of ONSM. Most ONSMs are tubular in shape, but some are globular or exhibit focal enlargement.¹⁵ The present patient had a tram-track pattern of enhancement on axial and coronal images of a tubular tumour looking more like an ONSD than an ONG (Figure 1).

The mechanism causing a CRVO in ON tumours is likely compression of the retinovascular bundle in the distal optic nerve, similar to occlusions in the setting of disc oedema of other etiologies.^{9,16,17} However, a CRVO has not been reported in a child with either an ONG or a ONSM. Retinochoroidal vessels may appear on the surface of the disc, often as disc swelling begins to resolve, and they are virtually pathognomonic of central retinal vein (CRV) compression. These vessels deliver venous blood from the retinal to the choroidal circulation, thereby bypassing the CRV and allowing retinal blood egress via the superior

or inferior ophthalmic veins. In our patient, retinochoroidal collaterals were visible in the presence of disc oedema. Retinal arteriovenous anastomoses were also present, another sign of defective retinal venous drainage.

Other orbital mass lesions may also cause optic disc oedema, including thyroid eye disease with intraorbital compression of the optic nerve,^{2,16} neurosarcoidosis,¹⁶ and optic nerve infiltration by malignant meningitis.² Examination and neuroimaging in this boy eliminated thyroid eye disease as a diagnostic possibility. He had no history of malignancy, and it is unlikely that malignant meningitis would cause no additional medical problems beyond optic nerve sheath infiltration over a 3-year period. The triad of visual loss, optic atrophy, and retinochoroidal collaterals was reported to be commonly caused by an optic nerve sheath meningioma,¹¹ but this observation is neither sensitive nor specific in the setting of paediatric patients.¹⁸

In summary, we describe the first reported paediatric patient with an aggressive optic nerve tumour of uncertain histology but suggestive of ONSM causing a CRVO, optociliary shunt vessels, and arteriovenous anastomoses.

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Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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