Multifocal malignant optic glioma of adulthood presenting as acute anterior optic neuropathy

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ABSTRACT

We report a 63-year-old, previously healthy female patient with glioblastoma multiforme of the optic nerve and chiasm presenting as acute anterior optic neuropathy. She presented with a 3-week history of progressively increasing headaches, retrobulbar pain, rapidly progressive visual loss in the right eye and blurred vision in the left eye. Early clinical examination revealed right optic disc swelling and she was initially diagnosed with demyelinating optic neuritis. Her clinical course deteriorated with total visual loss in the right eye and progressive visual loss in the left eye despite treatment with intravenous (IV) methylprednisone and IV immunoglobulins. MRI revealed enhancement of the right optic nerve and optic chiasm, with multiple periventricular hyperintense foci. Six weeks later, the patient presented with left facial palsy and left hemiparesis. Follow-up MRI showed multiple enhancing lesions in addition to the previous lesions involving right lentiform and right thalamic nucleus, right cerebral peduncle, right temporal and parietal lobes. Although the optic nerve biopsy was inconclusive, the brain biopsy revealed glioblastoma multiforme. This report demonstrated that malignant glioma of adulthood may be multicentric and may mimic optic neuritis clinically, which might help explain the difficulties in diagnosis.

1. Case report

A 63-year-old previously healthy woman presented with a 2-day history of rapidly progressive visual loss in the right eye associated with retrobulbar pain. Initial neuro-ophthalmic examination revealed best corrected visual acuity (VA) of hand movements (HM) in the right eye (oculus dexter [OD]) and 20/30 in the left eye (oculus sinister [OS]) with a right relative afferent pupillary defect (RAPD). The patient identified 0/15 OD and 14/15 OS Ishihara Colour Plates (ICP). Funduscopic examination showed a pale swollen right optic disc with no abnormality on the left. Erythrocyte sedimentation rate and C-reactive protein values were within normal limits and there were no other features of giant cell arteritis. A diagnosis of non- artertic anterior ischemic optic neuropathy was made.

Five days later the patient's vision deteriorated further with worsening headaches but no neurological or systemic symptoms. Visual acuity had deteriorated to perception of light OD and 20/50 OS, decreased ICP to 9/15 with upper temporal visual field loss OS.

Her MRI showed marked enlargement and enhancement of the intraorbital and intracranial parts of the right optic nerve and chiasm (Fig. 1a, b). There were also multiple, high signal foci in the periventricular deep white matter, parieto-occipital and fronto-parietal regions, right thalamus and right lentiform nucleus (Fig. 1c). Her spinal MRI was unremarkable. Lumbar puncture (LP) and extensive blood testing did not reveal any abnormalities except for mildly elevated serum antinuclear antibodies. The patient was admitted to the Neurology Department and a diagnosis of acute disseminated encephalomyelopathy was made. The patient was treated with 5 days of intravenous (IV) methylprednisolone with some subjective improvement in visual function. One week later her follow up examination revealed no light perception (NLP) OD, 20/30 and ICP 15/15 OS. Fundoscopic examination revealed multiple retinal haemorrhages, mildly engorged retinal veins and persistent disc oedema in the right eye (Fig. 2). No abnormalities were seen in the left eye. The patient was prescribed a tapering course of oral prednisone, which temporarily stabilised her vision.

One month later, the patient returned with weight loss of 7 kg to 10 kg, increasing headaches, hoarse voice and deteriorating left vision. Examination showed left facial weakness, NLP OD, 20/60 OS, decreased ICP to 9/15 and complete temporal hemifield loss. Funduscopy revealed a pale right optic disc and moderate temporal disc pallor in the left eye. She was again placed on a course of IV methylprednisolone with some improvement in her VA to 20/30 OS, 15/15 ICP, return of her normal voice, and improvement in facial weakness.

Repeat brain MRI showed persistent enhancement of the enlarged right optic nerve and chiasm with multifocal hyperintense lesions and expansion of the right lentiform lesion (Fig. 3a). A radiological diagnosis of multiple sclerosis was made. A few days later the patient's vision in the left eye deteriorated further and she was given IV immunoglobulins for 3 days without any improvement. A right optic nerve biopsy was inconclusive.

CT scans of the chest, abdomen and pelvis, total body bone scan, colonoscopy, mammogram, tumour markers, repeat LP and cerebral spinal fluid for cytology were all negative for malignancy.

The patient re-presented 1 week later with progressive visual deterioration in the left eye with VA OS 6/36 ICP 3/15, left facial weakness and left hemiparesis. The right eye remained NLP with amaurotic pupil. Repeat MRI showed persistent enhancement of the right optic nerve and chiasm (Fig. 4b). There was a heterogeneously enhancing mass within the right lentiform nucleus invading the posterior limb of the internal capsule with other smaller lesions involving: right cerebral peduncle, right parietal lobe, superior right temporal gyrus and right thalamic nucleus. There were multiple foci of high signal throughout the cerebral white matter, more confluent within the parietal lobe bilaterally (Figs 3b, c and 4a, c, d).

Her condition deteriorated further with NLP in both eyes and left hemiplegia. Brain biopsy performed through a right parietal burr hole showed multicentric glioblastoma multiforme. The patient was treated with chemotherapy (temozolomide) and was dis-

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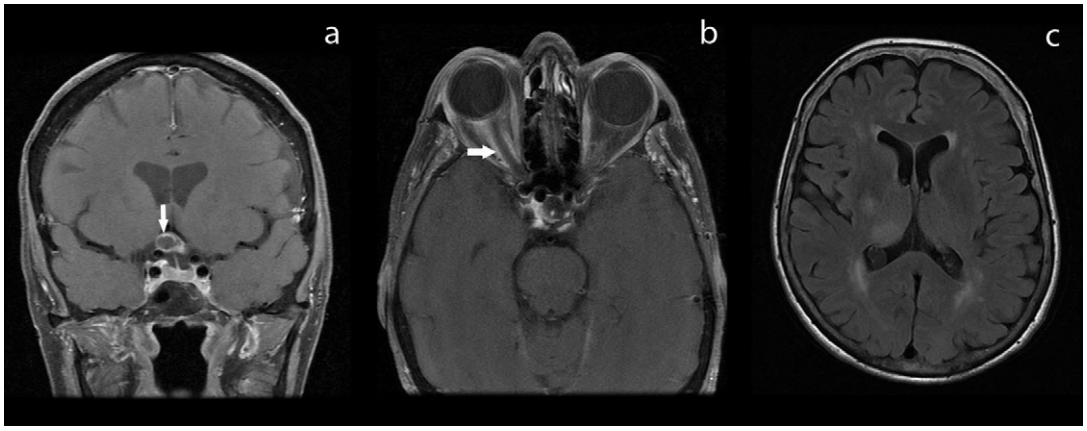

Fig. 1. (a) Coronal T1-weighted contrast-enhanced MRI showing ring enhancement of the chiasm (arrow); (b) axial T1-weighted contrast-enhanced MRI showing enlargement and enhancement of the right optic nerve (arrow); (c) axial fluid attenuated inversion recovery (FLAIR) MRI showing high signal in the periventricular deep white matter, multiple foci involving the parieto-occipital and fronto-parietal regions.

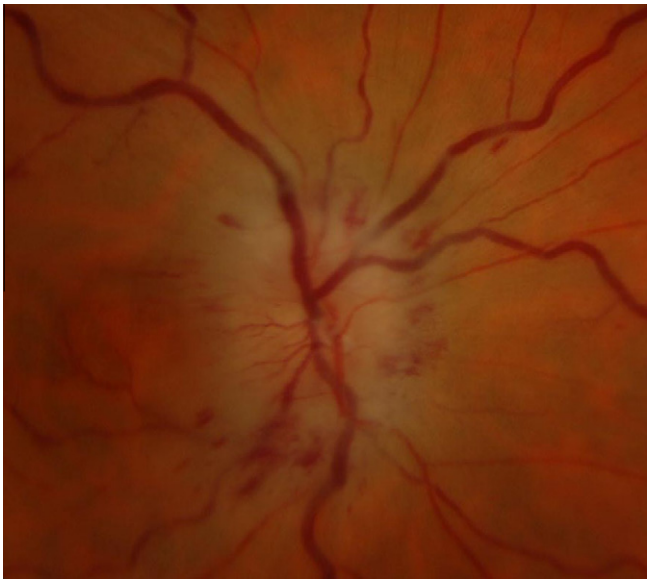

Fig. 2. Photograph of the fundus showing a swollen right optic disc with multiple retinal haemorrhages and venous engorgment. (This figure is available in colour at www.sciencedirect.com.)

charged to a hospital-based rest home. The patient died 6 months following her initial presentation.

2. Discussion

Optic nerve gliomas are rare, aggressive tumours in adults that are distinctly different from the more common optic gliomas of childhood. Hoyt et al.[1] described a "syndrome of malignant optic glioma of adulthood" in a landmark article in 1973. These authors identified four characteristic features: (i) presentation in middle-aged adult men; (ii) signs and symptoms mimicking optic neuritis; (iii) a progression to blindness within 5 weeks to 6 weeks; and (iv) death within several months.

Acute unilateral visual loss is the presenting symptom in 70% of patients with optic nerve gliomas and almost one-third have associated neurologic symptoms (headache, hemiplegia or dementia).[2] The specific pattern of visual loss depends on the site of origin of the tumour. Tumours that originate in the proximal portion of the nerve produce a characteristic monocular blurring of vision, optic nerve head swelling and periorbital pain. Subpial tumour growth produces venous obstruction, an appearance consistent with venous stasis retinopathy or central vein occlusion as described in our patient. When the origin is in the distal portion of the optic nerve or the optic chiasm, the visual loss may be simultaneously bilateral or nearly so and is associated with a pale or normal-appearing optic disc (posterior ischemic optic neuropathy).[3]

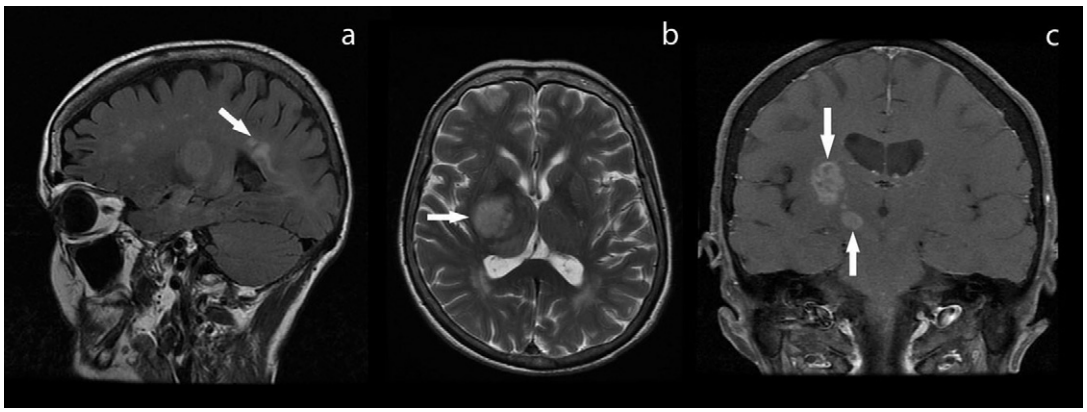

Fig. 3. (a) Sagittal fluid attenuated inversion recovery (FLAIR) MRI showing a large mass in the right lentiform nucleus with multiple hyperintense foci in the parieto-occipital and parieto-frontal regions (arrow); axial T2-weighted (b) and coronal T1-weighted contrast enhanced (c) MRI displaying enhancing lesions in the lentiform and thalamic nuclei (arrows).