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Case Report

Malignant optic glioma masked by suspected optic neuritis and central retinal vein occlusion

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ABSTRACT

Malignant optic glioma presents a clinical and diagnostic challenge, as early imaging findings overlap with other more common causes of optic nerve enhancement and enlargement, potentially leading to delay in diagnosis. This rare diagnosis carries an extremely poor prognosis, with death usually occurring within 1 year. We present a case of malignant optic glioma that was initially diagnosed as optic neuritis and central retinal vein occlusion, and we emphasize the importance of serial imaging and definitive biopsy to promote early diagnosis and treatment of this entity.

Introduction

Malignant optic gliomas (anaplastic astrocytoma or glioblastoma [GBM]) are very rare and most commonly present in older adults, without gender predilection. Prognosis for this condition is extremely poor, with death usually occurring within 1 year. Diagnosis of this entity can be difficult, as early imaging findings of malignant optic gliomas often overlap with other causes of unilateral optic nerve enlargement and enhancement such as ischemic optic neuropathy, optic neuritis, central retinal vein occlusion (CRVO), neurosarcoidosis, leukemia, lymphoma, low-grade optic nerve glioma, or metastasis [1,2]. We highlight the clinical and diagnostic challenge of this rare entity, especially when masked by a clinical presentation of optic neuritis and CRVO.

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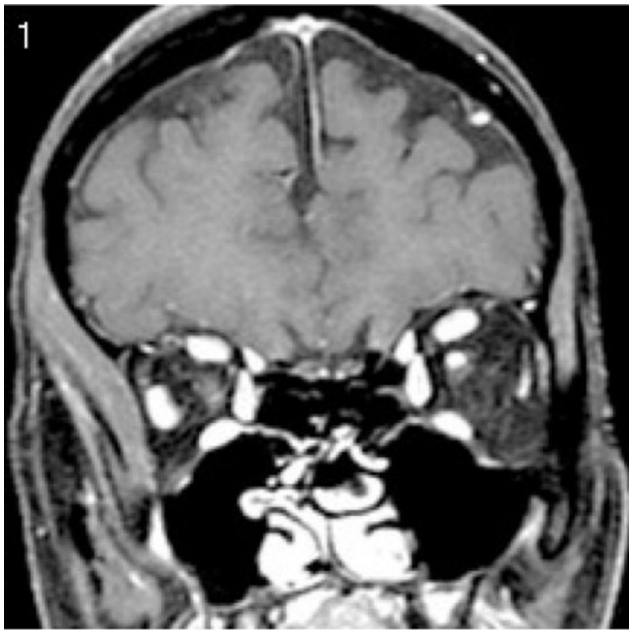

Fig. 1 – Coronal contrast-enhanced T1-weighted image of the orbits with fat saturation shows focal enhancement within the superior aspect of the left optic nerve.

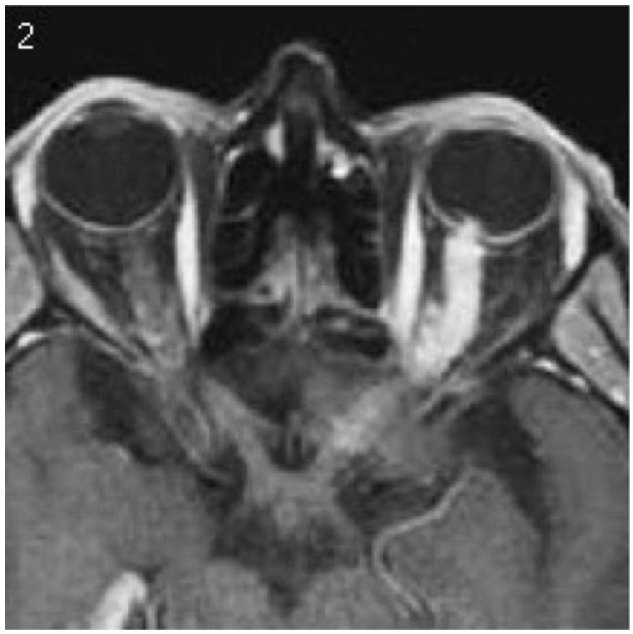

Fig. 2 – Axial contrast-enhanced T1-weighted image of the orbits with fat saturation 4 months after initial imaging shows abnormal enhancement and enlargement of the left optic nerve. Enhancement extends from the posterior globe to the distal prechiasmatic segment of the optic nerve.

Case report

A 72-year-old female with a history of Waldenstrom macroglobulinemia and prior irradiation of the left neck presented with new onset left orbital pain associated with eye movement and left inferior nasal quadrantanopia. The patient was evaluated by an ophthalmologist who confirmed swelling of the left optic disc and the presence of a visual field defect. The patient's symptoms and exam were felt to be most consistent with optic neuritis. An MRI of the orbits was performed and reported as normal. The patient was treated conservatively with analgesics.

The patient's left eye vision worsened over the next 3 months. Neuro-ophthalmologic evaluation led to the clinical diagnosis of CRVO. The initial MRI (Fig. 1) was rereviewed by the neuro-ophthalmologist, who retrospectively detected enhancement of the left optic nerve. This was thought to be related to the patient's CRVO, although the possibility of lymphoma of the optic nerve was considered. The patient was treated with intravitreal bevacizumab injection, laser panretinal photocoagulation, and oral acetazolamide.

One month later, the patient progressed to complete blindness on the left. Repeat MRI demonstrated findings of diffuse left optic nerve enhancement (Fig. 2). The patient was again evaluated by the neuro-ophthalmologist, who found no light perception in the left eye. The diffuse enhancement of the left optic nerve on MRI was attributed to progression of ischemic optic neuropathy following CRVO, although other potentially treatable conditions such as lymphoma, sarcoidosis, and optic neuritis could not be excluded. The patient was referred to the oculoplastic service for biopsy, and a follow-up MRI was ordered.

Repeat orbital MRI performed 3 weeks later (Fig. 3) showed increased enlargement and enhancement of the left optic nerve as well as increased perioptic stranding. Imaging findings were still thought to be nonspecific, with differential considerations including optic neuritis, lymphoproliferative disease, primary glioma, and infection. Given the uncertainty in diagnosis, strong clinical suspicion for an inflammatory and ischemic process, and MRI features of some optic nerve sheath and perioptic infiltration, the patient underwent a biopsy of the optic sheath. Histopathologic examination revealed no evidence of malignancy or inflammation.

Two months later, the patient noted new right superior temporal quadrantanopia. An urgent MRI was ordered and demonstrated progression of left optic nerve expansion and enhancement, with areas of necrosis and involvement of the optic chiasm (Fig. 4). These findings were considered to be highly concerning for malignant optic glioma, and the patient underwent biopsy of the optic nerve itself with pathology confirming GBM.

Despite treatment with chemoradiation, the patient's right vision loss worsened. The patient ultimately expired 15 months from initial onset of symptoms due to ischemic colitis, a complication of Waldenstrom's macroglobulinemia.

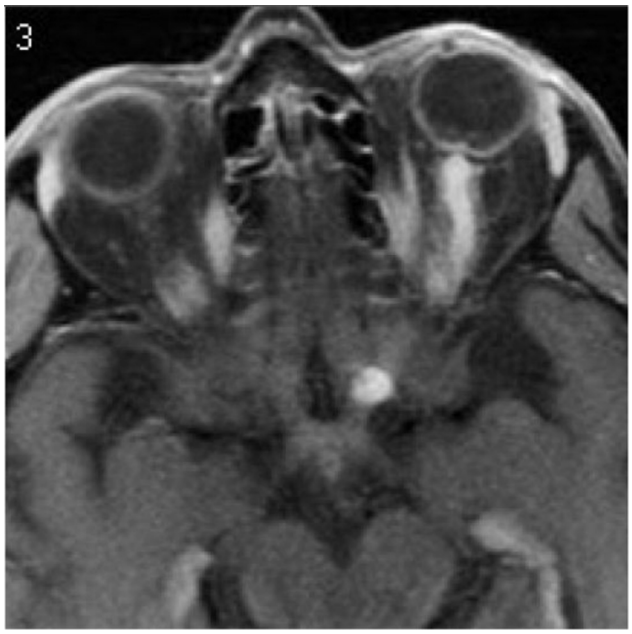

Fig. 3 – Axial contrast-enhanced T1-weighted image of the orbits with fat saturation 5 months after initial imaging shows increased extent of enhancement of the left optic nerve, which now appears somewhat nodular in the prechiasmatic segment.

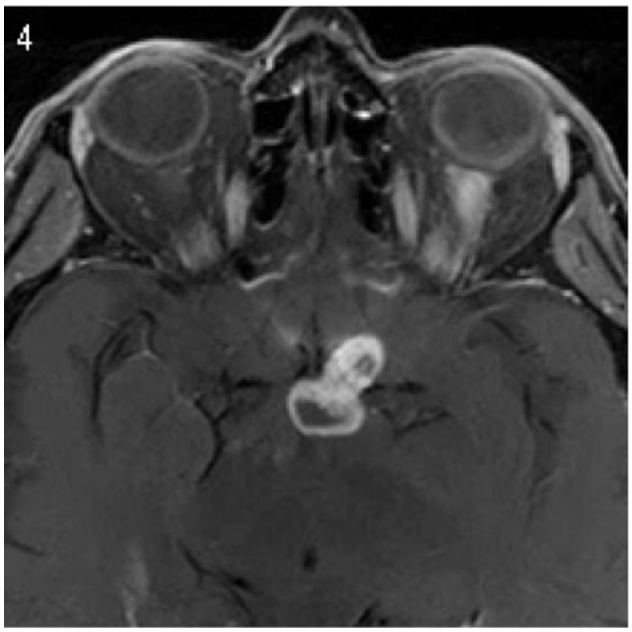

Fig. 4 – Axial contrast-enhanced T1-weighted image of the orbits with fat saturation 7 months after initial imaging shows marked progression of disease with increased size, mass-like enhancement, and necrosis of the prechiasmatic left optic nerve lesion and the optic chiasm.

Discussion

Malignant optic gliomas of adulthood are classified pathologically as either an anaplastic astrocytoma or a GBM multiforme, are very rare, and are more commonly present in the second to eighth decades of life, with no definite gender predilection [1,3-7]. The clinical presentation of malignant optic gliomas of adulthood typically begins with visual field deficits that relate to the location of tumor, followed by rapidly progressive vision loss, with this progression often worsened by chiasmatic involvement [1,3,4,7,8].

The diagnostic challenge in our patient centered on the initial presentation, as her clinical examination findings of monocular vision blurring and retrobulbar pain overlapped with the clinical diagnosis of optic neuritis and her clinical examination findings were also compatible with ischemic CRVO. Even when imaging is performed to help narrow the differential diagnosis, early imaging findings of malignant optic nerve gliomas, including enhancement and enlargement of the optic nerve, are often nonspecific and may overlap with optic neuritis, CRVO, or other entities [1,2]. While the initial clinical presentation and imaging of malignant optic nerve gliomas are nonspecific, the pathologic findings are characteristic. Precise diagnosis depends on biopsy findings, but a biopsy of the optic nerve is risky and only undertaken when there is a high index of suspicion for an aggressive lesion [1]. Nevertheless, biopsy of the optic nerve itself and not the optic nerve sheath should be undertaken, if there is a strong suspicion for malignant optic nerve glioma.

Treatment of malignant optic nerve gliomas may include radiation, chemotherapy, or both; a combination of radiotherapy and temozolomide has been shown to increase median survival by 2-3 months [1,9]. Despite treatment, however, malignant optic gliomas typically carry a poor prognosis, with complete blindness within 5-6 weeks of initial presentation and death occurring within approximately 1 year [1,3,4,7-9].

The patient in this case exemplifies the diagnostic challenge presented by this rare entity given overlapping clinical symptoms with the far more common entities of optic neuritis and CRVO. This case demonstrates the additional challenge of early radiologic diagnosis of malignant optic glioma, as imaging findings are often nonspecific and can overlap with other causes of unilateral optic nerve enlargement and enhancement, including optic neuritis, ischemic optic neuropathy, central retinal vein occlusion, and infiltrative disorders such as neurosarcoidosis and lymphoma [3,10-12].

Short-term follow-up contrast-enhanced MR imaging of the orbits should be considered in such patients in order to evaluate for resolution of abnormal optic nerve enhancement, which would be expected in treated optic neuritis or evolution of CRVO, or to confirm persistent or progressive optic nerve enhancement as is seen in malignant optic nerve gliomas. Ultimately, confirmation of the diagnosis will depend on the combination of clinical presentation, response to treatment, biopsy, and serial imaging findings [13]. In this case, the focal nature of the initial enhancement as well as the rapid progression and extent of nerve enlargement were important clues to the diagnosis of malignant optic glioma.

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