CASE REPORT

Progressive optic nerve glioma: orbital biopsy technique using a surgical navigation system

An 11-year-old male child presented with worsening vision in his left eye. Examination and imaging revealed a left optic nerve tumour causing mass effect and optic neuropathy, without systemic evidence of neurofibromatosis. In view of the significant risk to visual acuity, a biopsy was deferred and chemotherapy was commenced. After initial stability, continued visual decline necessitated incisional biopsy. Surgical navigation was used to facilitate minimal access surgery avoiding bone removal. The system also precluded biopsy of cystic parts of the tumour, allowing successful intraoperative frozen-section confirmation of lesional tissue. Our case report serves to highlight specific circumstances where surgical navigation may be a useful tool for the orbital surgeon.

Case Report

An 11-year-old male child presented with a 3-month history of blurred vision in his left eye. He had no other visual or systemic symptoms. His medical history was unremarkable. He had an ocular history of bilateral optic disc drusen (Fig. 1A) that had remained stable with no effect on visual acuity or optic nerve function on previous reviews. On this examination, his vision was 20/20 right eye and 20/50 left eye. He had loss of color vision and an inferior visual field defect in the left eye on automated testing. He also had a left relative afferent pupillary defect with 2 mm of proptosis by Hertel exophthalmometery. Fundus examination showed severe edema of the left optic nerve (Fig. 1B). An urgent magnetic resonance imaging (MRI) was performed on the day of presentation and revealed a heterogeneously enhancing mass involving the intraorbital left optic nerve with minimal extension to the intracanalicular and prechiasmatic segments (Fig. 2A). Based on typical imaging features, a presumptive diagnosis of optic nerve glioma was made. Orbital biopsy was discussed between the patient, parents, and multidisciplinary team, but deferred because of the significant risk of iatrogenic worsening of vision in the affected eye.

The patient was started on vinblastine and followed closely. For 5 months, the patient's visual acuity and field remained stable and the optic nerve edema steadily improved. Six months after starting treatment, the patient noted a decrease in his vision. Examination revealed a 2-line loss of visual acuity, worsening of the visual field loss, and recurrence of the optic nerve edema. There was enlargement of the tumour on MRI with more posterior extension. The chemotherapeutic regimen was changed to vincristine and carboplatin.

In the 3 months after the change of chemotherapy, the patient's visual function continued to progressively decline, eventually reaching 20/1250, with severe, generalized loss of visual field. Repeat MRI showed a significant increase in the size of the prechiasmatic/chiasmatic component of the tumour with patchy T2/FLAIR nonenhancing hyperintensities extending into the bilateral optic tracts, hypothalamus, and internal capsules (Fig. 2B).

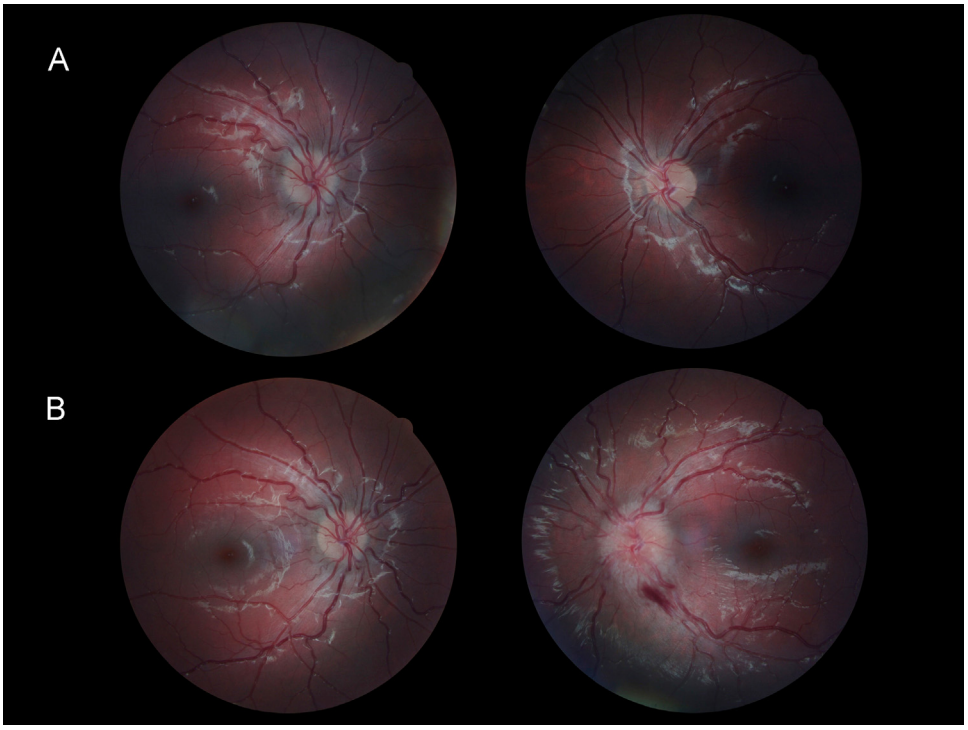

Fig. 1—(A) Fundal view at past follow-up with bilateral optic disc drusen. (B) Fundal view at initial presentation with decreased vision OS. Note significant disc edema, vessel blurring, and inferior flame-shaped hemorrhage.

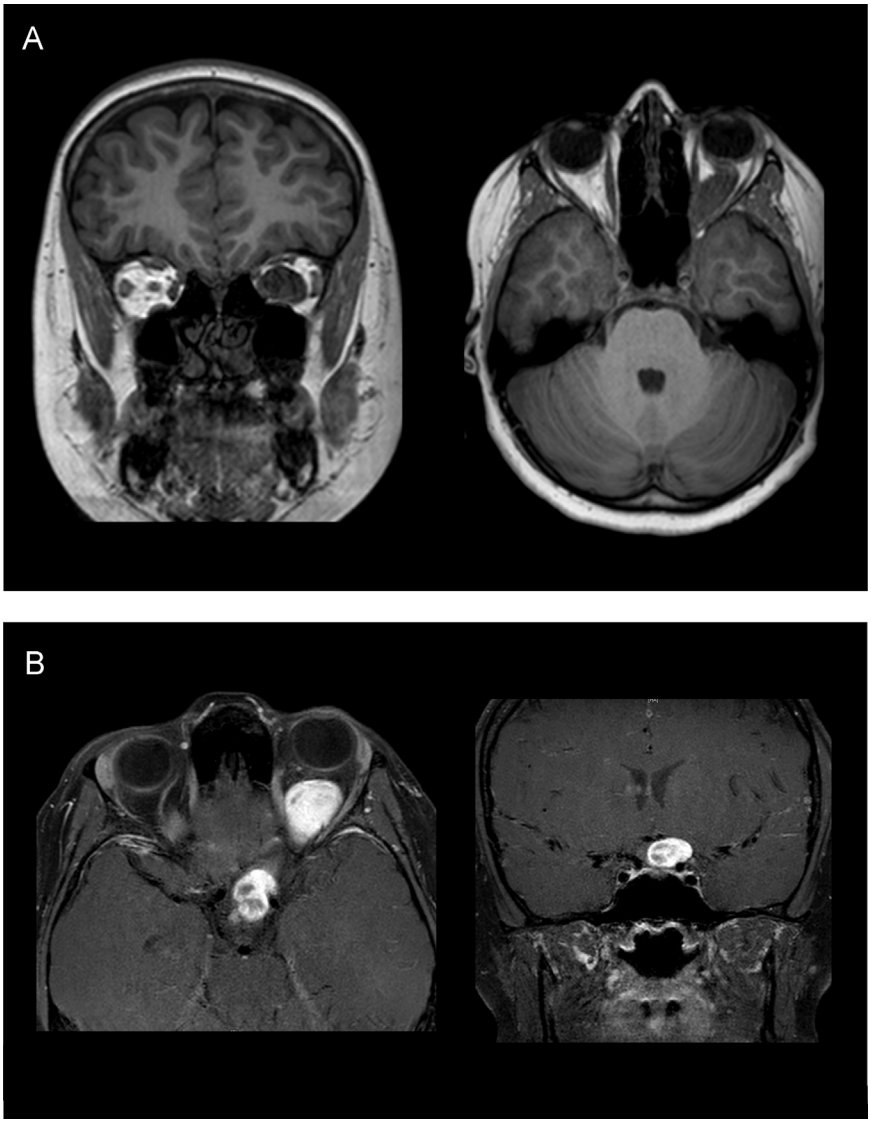

Fig. 2—(A) Magnetic resonance imaging (MRI) T1 coronal and axial views at presentation demonstrating intraconal tumour involving the optic nerve OS. (B) MRI T2 with contrast, axial, and coronal views 9 months after the start of chemotherapy demonstrating tumour growth and posterior extension.

The patient also developed a temporal visual field defect in the previously unaffected right eye.

On account of the worsening of visual function and enlargement on MRI, an orbital biopsy of the tumour was arranged. This was performed through a transconjunctival medial approach via an intraconal route; however, it was nondiagnostic as the tissue sample was found to contain only thickened dura with no lesional tissue. A further multidisciplinary meeting, including neurosurgery, oncology, and ophthalmology, concluded that a combined-approach tumour resection would present considerably morbidity to the patient and risk vision damage in the contralateral eye. It was agreed that a diagnostic incisional biopsy via a lateral orbitotomy with bone removal would allow further management options to be considered. At this point, consideration was given to the use of surgical navigation available within the unit.

The Medtronic Stealthstation S7® Surgical Navigation System uses an electromagnet placed in close proximity to the site of surgery that calibrates using a tracker in relation to preuploaded patient computed tomography (CT) and

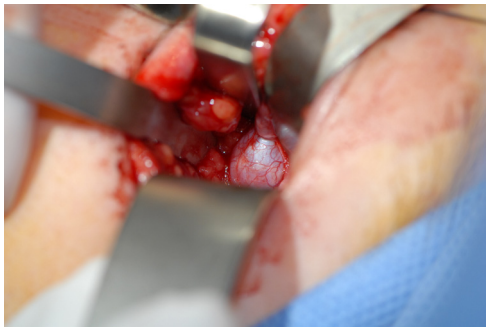

Fig. 3—Intraoperative view of tumour prebiopsy.