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CASE REPORT

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Surgical reversal of prolonged blindness from a metastatic neuroblastoma

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Abstract *Background:* Reports of tumor-related anterior visual pathway blindness that have resolved after surgical decompression are rare. The longest reported duration of tumor-related blindness completely reversed by optic nerve decompression is 3 days. We describe a pediatric patient with 7 days of no light perception who experienced reversal of blindness following tumor resection and optic nerve decompression. *Case description:* A 33-month-old boy presented with a 4-day history of no light perception. Magnetic resonance imaging revealed a mass involving the sphenoid sinus, sella turcica, and clivus with significant optic nerve involvement. Loss of light perception and complete absence of a pupillary light reflex were documented for the next 72 h. A sluggish pupillary light reflex was regained 24 h after instituting intravenous steroids. An urgent bi-frontal craniotomy and optic nerve decompression were performed 7 days after the onset of blindness. Surgical pathology revealed metastatic neuroblastoma. Eleven days after optic nerve decompression, the child was able to count fingers and recognize faces and printed book characters. *Conclusion:* Prolonged blindness secondary to tumor-related optic nerve compression may be reversible up to 1 week from onset in children presenting with no light perception.

Keywords Blindness · Neuroblastoma · Optic nerve · Surgical decompression · Visual pathways

Introduction

Acute onset of blindness due to optic nerve compression is most commonly the result of acute mass effect [12, 14] caused by pituitary apoplexy, cerebral artery aneurysm rupture, or trauma [15]. Although progressive tumor impingement on the optic nerve can cause visual field deficits, reports of bilateral anterior visual pathway blindness not caused by hemorrhage into a tumor are rare. To date, surgical reversal of tumor-related blindness has been limited to cases of pituitary apoplexy, meningioma, and craniopharyngioma [1, 7, 8, 10, 11, 16]; blindness from a compressive neuroblastoma has been considered "irreversible" [2]. We report the first case of prolonged blindness due to optic nerve compression by a neuroblastoma that has been reversed by decompression.

Case report

History and examination

A 33-month-old boy with no prior medical history presented with a 4-day history of vision loss. Neurological examination revealed complete loss of light perception and an absent pupillary constriction to light in both eyes. Fundoscopic examination showed bilateral optic disk pallor. No other physical abnormalities were noted. Computed tomography (CT) scan revealed a midline mass centered at the level of the clinoid extending into the ethmoid and filling the nasopharynx with no evidence of acute hemorrhage (Fig. 1). Magnetic resonance imaging (MRI) demonstrated a 36-cm^3 extra-axial, heterogeneous, poorly defined midline mass involving the clivus, ethmoid and

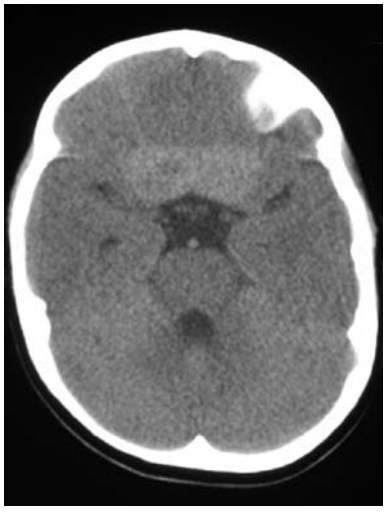

Fig. 1 Preoperative computed tomography scan of a 33-month-old male presenting with 4-day history of no light perception. A bilobed, midline, contrast-enhancing mass is centered at the level of the anterior clinoids, and extends into the ethmoid and nasopharynx with no evidence of acute hemorrhage.

sphenoid sinuses, lesser sphenoid wings, and sella turcica (Fig. 2). The optic nerves could not be identified at the foramen secondary to significant compression. A full-body MRI revealed a 3×2-cm mass involving the left adrenal gland, and multiple gadolinium-enhancing lesions within the cervical, thoracic, and lumbar vertebral bodies.

Treatment and subsequent course

An endoscopic ethmoidotomy with biopsy of the nasal and sphenoid sinus mass was performed. Microscopic examination revealed a poorly differentiated neuroblastoma. Twenty-four hours after instituting intravenous steroid therapy (7 days after the reported onset of blindness), the right pupil had a sluggish constriction to light (from 6 to 5 mm); however, the patient continued to have no light perception. The patient was immediately taken to the operating room for a bilateral frontal craniotomy, tumor debulking, and bilateral optic nerve decompression. Intraoperatively, both optic nerves were encased in tumor. Postoperative imaging demonstrated a partial resection of the mass along the anterior cranial fossa with decompression and clear visualization of both optic nerves and the chiasm (Fig. 3). On postoperative day 2, both pupils were briskly constricting (6 to 3 mm) in response to light. Perception of light returned on postoperative day 5. After 1 week, the patient could count fingers 2 ft away and tracked objects with his eyes. After receiving a 3-day course of vincristine sulfate, adriamycin, and cytoxan therapy, the patient was discharged home 11 days after surgery with the ability to recognize television and book characters.

Intervention

Following a bicoronal skin incision, an osteoplastic bifrontal craniotomy and a bilateral orbital bar osteotomy were performed. The pterional boney resection was extended posteriorly toward the orbital apex with Leksell rongeurs. After opening the dura, self-retaining retractors were used to gently elevate the frontal lobe, and the operating microscope was inserted to expose the opticocarotid triangle. The dura was abnormally elevated along the sphenoid wing by the tumor, which initially obscured the proximal intracranial carotid artery and the optic nerve. Additional exposure was achieved by opening the proximal sylvian fissure. Tumor growth within the dura had expanded and softened the anterior clinoid. An H-shaped incision into the dura overlying this area revealed firm, vascular, and somewhat friable tissue. Using sharp dissection under high microscopic magnification, we excised the clinoidal dura and removed the tumor overlying the distal dural ring, expanding the clinoid. The tumor also had eroded the optic canal, so we used a high-speed drill

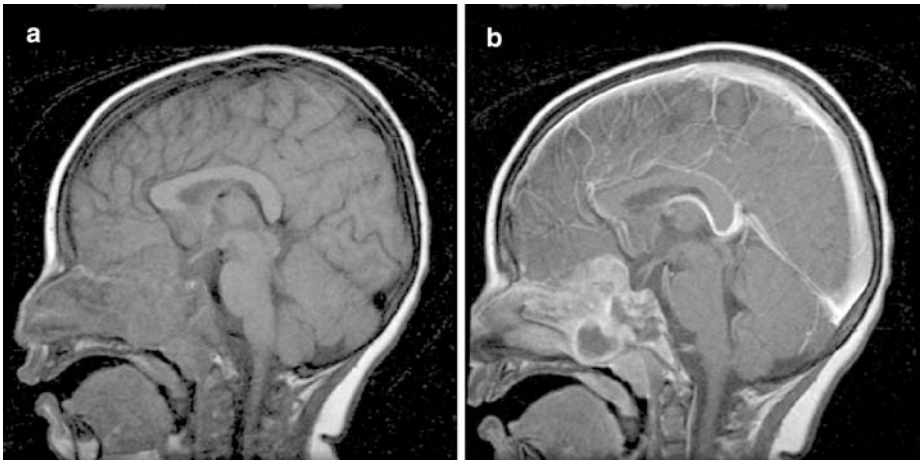

Fig. 2 Sagittal T1-weighted magnetic resonance images of a heterogeneous and poorly-defined mass centered at a level of the clinoid. **a** Pregadolinium. **b** Postgadolinium.

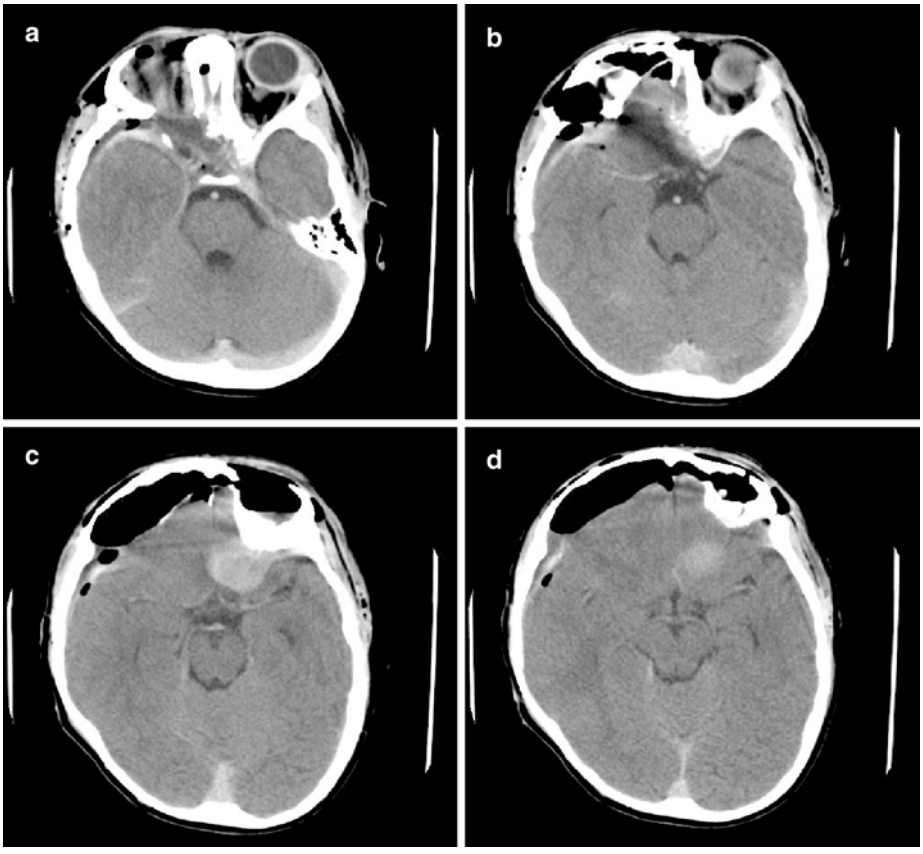

Fig. 3 Postoperative computed tomography scans revealing serial sections (**a**–**d**, inferior to superior) through the anterior skull base. The patient underwent a partial resection of the tumor with decompression of both optic nerves.

with a 3-mm diamond drill bit to remove residual bits of the eroded optic nerve canal and further release the optic nerve on the right side. The falciform ligament was also opened over the optic nerve, which had been bowed and stretched by the tumor. We opened the dural ring of the optic nerve and dissected the surrounding tissues and associated tumor until the nerve was entirely exposed to its entrance into periorbital tissues.

The tumor resection was then carried out medially into what would ordinarily be the region of the ethmoid sinuses but which, in this patient, had been entirely displaced by tumor. Additional layers of bone were removed until the left optic nerve was identified and decompressed in like fashion.

Closure was accomplished utilizing a dural graft where the tumor had been excised, and by rotating in a vascularized pericranial graft overlying the region of the ethmoids (to preclude omission of any open sinus areas). The orbital bar and the osteoplastic craniotomy flap were then reattached with absorbable microplates and screws.

Discussion

Anterior visual pathway compression blindness has previously been described as reversible within 72 h of onset [1, 5, 7, 8, 10, 11, 16–18]. Al-Wahhabi et al. reported reversal of 72-h blindness following optic nerve decompression in a 3-year-old with a craniopharyngioma [1]. Tajima et al. described reversal of blindness following decompression 48 h after loss of vision in a patient with a large unruptured anterior cerebral artery aneurysm [18]. Optic nerve decompression in the setting of prolonged blindness has been less encouraging, however. Maurer et al. reported no improvement in vision following optic nerve decompression in 7 patients experiencing blindness for 4 or more days due to a meningioma [11]. Other authors have also reported failure to reverse meningioma-induced blindness with optic nerve decompression when loss of light perception was present for more than 1 week [8, 10, 16]. Striph et al. reported the only case of reversed blindness greater than 72 h after onset, accomplished by decompressing a giant anterior communicating artery aneurysm 5 days after loss of light perception [17].

The underlying pathophysiology of optic nerve compression is poorly understood. Ischemic mechanisms are the most widely accepted cause of optic nerve dysfunction following compression [6]. In our case, the 10-day duration of recovery as well as the near-complete return of vision would argue against vascular compromise caused by arterial insufficiency [3] or venous stasis [9]. Clifford-Jones et al. demonstrated that optic nerve demyelination occurs within 1 week of compression, with remyelination beginning after 5 weeks of compression [4]. It is unclear

how the more rapid and near-complete recovery made in our patient could occur by this more chronic mechanism. Stretching of the axonal membranes or deformation of myelin with displacement of the nodes of Ranvier [13] allows for a reversible conduction block in subacute compression of the optic nerve and may explain the phenomenon observed in our patient.

Despite complete blindness from optic nerve compression, our patient demonstrates that if some discernible pupillary light reflex is present, surgical decompression can result in very good recovery of vision, even when delayed for longer than 72 h.

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