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A Review of Visual and Oculomotor Outcomes in Children With Posterior Fossa Tumors

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Tumors of the posterior fossa represent the most common solid malignancy of childhood and can affect the visual system in several ways. This article outlines the relevant visual anatomy affected by these tumors and reviews the visual and oculomotor outcomes associated with the following 3 most common tumor types—medulloblastoma, juvenile pilocytic astrocytoma, and ependymoma. The available data suggest that the rate of permanent vision loss is low (5.9%-8.3%), with patients having juvenile pilocytic astrocytoma demonstrating the best outcomes. The rate of long-term strabismus (25%-29.1%) and nystagmus (12.5%-18%) is higher and associated with significant morbidity.
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Introduction

Pediatric brain tumors represent the most common solid malignancy of childhood, and visual dysfunction is cited as the most frequent presenting symptom after headache and vomiting.[1-5]

Tumors of the posterior cranial fossa, the region of the brain most often involved in the pediatric population, may affect visual function in several ways. Compression or infiltration of the normal cerebrospinal fluid (CSF) outflow pathways can lead to hydrocephalus and papilledema, which is reported to occur in up to half of all patients presenting with a posterior fossa mass.[6-9] Swelling at the optic nerve head is associated with varying degrees of visual acuity loss. Although papilledema typically resolves following treatment either by surgical resection of the tumor or neurosurgical shunting for hydrocephalus, severe or prolonged swelling of the optic nerve can result in axonal death, optic atrophy, and permanent vision loss. Tumors involving the cerebellum and brainstem may also lead to oculomotor dysfunction, resulting in nystagmus and strabismus.[10,11]

The purpose of this article is to outline the relevant visual anatomy affected by tumors of the posterior fossa and review the visual and oculomotor outcomes associated with the three most common tumor types.

Relevant Anatomy

Papilledema (Fig. 1) refers to optic nerve head swelling secondary to increased intracranial pressure (ICP) (ie, a lumbar puncture opening pressure of more than 25-28 cm H_2O in children).[12,13] High pressure is transmitted to the fluid spaces around the optic nerve and leads to axoplasmic stasis and swelling at the optic nerve head.[14] The swollen nerve fibers can then compress the capillaries and venules on the optic disc, causing venous dilation and peripapillary hemorrhages. Venous stasis may also lead to disappearance of normal spontaneous venous pulsations in the larger venuoles overlying the optic disc.[15,16]

In the setting of posterior fossa neoplasms, the etiology of increased ICP and papilledema is obstructive (noncommunicating) hydrocephalus. Normally, CSF flows from the lateral ventricles to the third ventricle via the foramina of Monroe, and then through the sylvian aqueduct to the fourth ventricle in the brainstem (Fig. 2). From here, CSF can either pass into the central canal of the spinal cord or into the cisterns of the subarachnoid space. Tumors of the posterior fossa (ie, those arising from the brainstem or cerebellum) tend to compress or obstruct the sylvian aqueduct or fourth ventricle, leading to

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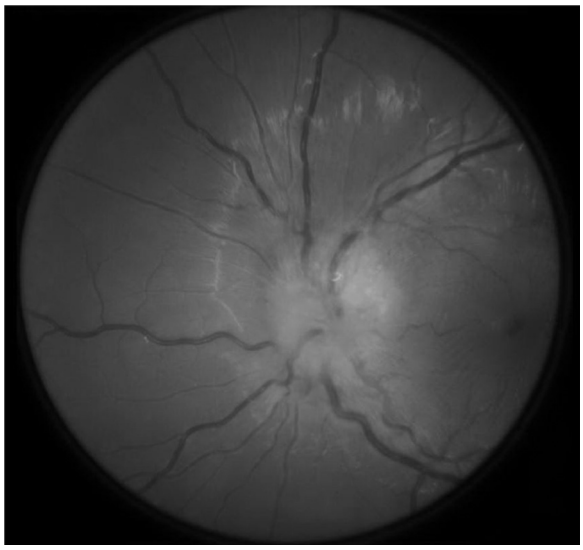

Figure 1 Swelling of the optic nerve secondary to increased intracranial pressure, with characteristic blurring of the optic disc margin and obscuration of the overlying vessels.

hydrocephalus. Typically, elevated ICP must be present for at least 1-5 days before papilledema develops.[17] In very young infants, papilledema may not develop at all if the cranial sutures have not yet fused and the cranial vault is able to expand in response to the increased ICP.[18]

The vision changes associated with papilledema range from mild to severe and are related to dysfunction at the optic nerve head.[19] One of the earliest changes that may be perceived by the patient (and can be detected on formal visual field testing) is enlargement of the physiological blindspot. This is primarily due to a mechanical or structural change caused by displacement of the peripapillary retina, resulting in a hyperopic shift around the optic nerve.[20] More significant swelling of the optic nerve may cause nasal, arcuate, or central visual field loss related more directly to axonal dysfunction. In these cases, a combination of axoplasmic stasis and ischemia likely

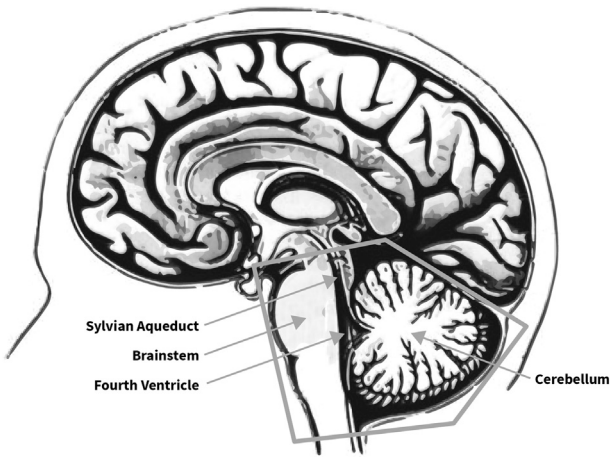

Figure 2 Borders of the posterior cranial fossa (thick gray line), located between the tentorium cerebelli (superiorly) and foramen magnum (inferiorly). Tumors arising from the brainstem or cerebellum can obstruct the sylvian aqueduct or fourth ventricle, causing hydrocephalus and papilledema.

contribute to the vision loss.[21] Fortunately, improvement in ICP from surgical debulking or CSF shunting procedures tends to result in rapid improvement in vision. Prolonged or severe papilledema can lead to axonal death, however, with subsequent optic nerve atrophy and permanent vision loss.[22]

Whereas optic nerve dysfunction is an indirect consequence of posterior fossa tumor growth (ie, the result of increased ICP), oculomotor issues often arise from direct tumor infiltration or compression of the pons, midbrain, and cerebellum. Several series report posterior fossa tumors as the most common cause of sixth nerve palsies in children, related to infiltration or compression of the sixth nerve nucleus in the dorsal pons.[23-25] Similar compression of midline structure can cause horizontal gaze palsies or, in the case of midbrain involvement, vertical eye movement abnormalities. As with papilledema, sixth nerve palsy can also result from increased ICP when downward displacement of the brainstem exerts a tractional force on the sixth nerve as it enters Dorello's canal.

Many of the circuits necessary for gaze holding are also found in the brainstem and cerebellum and tumor-related dysfunction in these areas can lead to nystagmus, an involuntary, rhythmic oscillation of the eyes. Periodic alternating, downbeat, upbeat, and torsional nystagmus are the forms that most commonly localize to lesions in the posterior fossa.[26-29] The degree to which these forms of oculomotor dysfunction improve following treatment depend on the tumor's growth pattern (compressive vs infiltrative) and the surgical options available, as discussed later.

Medulloblastoma

Medulloblastoma (MB), a tumor of embryonal origin, is one of the most common posterior fossa neoplasm in children with an annual incidence of around 6 per million. Tumors initially arise in the cerebellum, though metastases to the thoracic and lumbosacral spine occur in up to 40% of patients. Despite the risk of local metastasis, current treatment protocols that include surgical resection, craniospinal radiation, and chemotherapy can achieve 10-year survival rates of approximately 63%.[30-32]

The site of origin in the cerebellum puts patients with MB at risk for hydrocephalus, increased ICP, and papilledema as the tumor often compresses the fourth ventricle and sylvian aqueduct. An emergency external ventricular shunt may be required at the time of presentation and up to 30% of patients may need permanent ventriculo-peritoneal shunting following surgical resection of the tumor due to scarring of the CSF outflow pathways.[30] Compression, infiltration, and surgical manipulation of the cerebellum and brainstem may also cause oculomotor dysfunction.

Though outcomes data in the literature are limited, a UK study by Cassidy et al[33] documented the long-term visual and oculomotor sequelae of 24 pediatric MB survivors. All patients included in the study had undergone surgical resection of their tumor, all patients 3 years of age or older received craniospinal irradiation, and 17 children (70.1%) were treated with adjuvant chemotherapy. At the time of tumor diagnosis, 17