Atypical external hydrocephalus with visual failure due to occult leptomeningeal dissemination of a pontine glioma

Case report

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The authors report on the case of a diffuse pontine glioma in a 5-year-old boy in whom radiologically and cytologically occult leptomeningeal metastases led to the development of an atypical "external" hydrocephalus, associated with grossly elevated intracranial pressure (ICP). Initial neuroimaging demonstrated only mild communicating ventricular dilation associated with a noticeable enlargement of the subarachnoid space, particularly over the surface of the frontal lobes; these features are not usually associated with significantly elevated ICP. Possible pathophysiological mechanisms resulting in this unusual clinical presentation are discussed. Early recognition of the severity of the raised ICP despite the paucity of clinical and radiological signs may have averted the development of blindness due to optic atrophy.

Key Words • glioblastoma • radiotherapy • optic atrophy • cerebral atrophy • cerebrospinal fluid shunt • leptomeningeal gliomatosis

Brainstem tumors account for up to 25% of all intracranial neoplasms of childhood, of which 60 to 80% are diffuse intrinsic brainstem gliomas. Diffuse pontine gliomas occur most frequently in children between 6 and 10 years of age. The tumors are usually characterized by an aggressive clinical course and the patient typically dies within 6 to 12 months of diagnosis. Radiotherapy has been shown to be effective in extending patient survival but the benefit is small; chemotherapy does not influence disease progression. Multiple, often bilateral, cranial nerve involvement, ataxia, and long-tract signs are common clinical findings at presentation. In contrast, the finding of raised ICP at presentation is exceptional. Hydrocephalus usually arises secondary to aqueductal obstruction by the expanding tumor mass and generally occurs late in the disease course. A single case report of an adult who developed a communicating hydrocephalus secondary to leptomeningeal metastasis from an occult pontine glioma has been reported.

We present the unusual case of a diffuse pontine glioma in a child in whom early papilledema was found at presentation. The absence of radiological evidence of ventriculomegaly or tumor dissemination on initial imaging led to a delay in the recognition of the development of raised ICP, which resulted in blindness due to optic atrophy. Possible pathogenic mechanisms leading to this unusual presentation are discussed together with recommendations for the identification and management of similar cases.

Case report

History and Examination. This 5-year-old boy presented to an ophthalmologist with a 2-day history of diplopia and bilateral mild papilledema. Visual acuity was 20/20 in both eyes. There were no other abnormal neurological findings. An MR image was obtained (Fig. 1) that demonstrated a diffuse, minimally enhancing, pontine lesion extending superiorly into the midbrain, with no evidence of hydrocephalus (Fig. 1 D–F).

Initial Treatment. Radiological appearances were typical for a pontine glioma and, in accordance with current recommendations, the child was referred directly for radiotherapy without undergoing biopsy sampling of the lesion. Two weeks after commencing a 6-week course of fractionated radiotherapy, right arm weakness, bilateral ptosis, and bilateral papilledema developed. A diagnosis of radiation-induced brainstem edema was made. The symptoms completely resolved within 24 hours of commencing radiotherapy.

Posttreatment Course. Six weeks later, he presented again after a generalized tonic-clonic seizure followed by a transient episode of dysphasia, which was attributed to radiation-induced cerebral atrophy. Three weeks later, 7 months after his initial presentation, he was referred for a lumbar puncture. A diagnosis of raised ICP based on clinical examination and aque ductal pressure testing was made. The symptoms completely resolved within 24 hours of commencing radiotherapy.

Discussion

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Pontine glioma with external hydrocephalus

J. Neurosurg: Pediatrics / Volume 102 / March, 2005

102:224–227, 2005

A 9-year-old boy presented with a 6-month history of diplopia and gait ataxia. There was a partial left oculomotor nerve palsy with associated hemorrhagic papilledema on fundoscopy. Magnetic resonance imaging was performed (Fig. 2), which demonstrated significant shrinkage of the pontine tumor mass in response to the radiotherapy. The Sylvian aqueduct was patent and all four ventricles were mildly dilated with a noticeable enlargement of the subarachnoid space, particularly over the surface of the frontal lobes. The appearances of the ventricles were similar to those seen in external hydrocephalus of infancy, although the prominent widening of the perisylvian CSF spaces was somewhat atypical. Appearances were also compatible with a diagnosis of radiation-induced cerebral atrophy. The biopsy sampling confirmed a histological diagnosis of glioblastoma multiforme. Intraoperative CSF cytological examination also demonstrated the presence of malignant cells. The child was referred for palliative care and died 8 weeks later, 7 months after his initial presentation.
show an uncomplicated diffuse pontine glioma, and the early papilledema was erroneously attributed to the tumor mass. The insidious development of visual failure preceded the development of significant headache, which occurred late in the clinical course. Imaging studies at the time of visual loss were also misleading as only mild ventriculomegaly and obliteration of the subarachnoid spaces over the surface of the frontal lobes. These appearances were strikingly similar to those seen in benign external hydrocephalus, a self-limiting condition that is not usually associated with significant elevations of ICP.1,4 The development of hydrocephalus in this case was almost certainly secondary to obstruction of the pathways for CSF absorption by the widespread leptomeningeal metastases. Leptomeningeal metastases occur in up to one third of children with malignant pontine glioma. They may be radiologically and cytologically occult at presentation; repeated neuroradiological imaging and CSF sampling (both lumbar and ventricular CSF) may be required to establish the diagnosis.3,10,20

Typically, leptomeningeal metastases would be expected to result in a communicating hydrocephalus with progressive ventriculomegaly and obliteration of the subarachnoid spaces over the surface of the brain.4 For an external hydrocephalus to develop other pathological factors must be present. The lack of ventricular enlargement in this case probably reflects a relative loss of ventricular compliance due to subependymal tumor spread in the walls of the lateral ventricles. Although pathological confirmation for this hypothesis could not be obtained, MR imaging, obtained late in the clinical course, clearly demonstrated evidence of multiple subependymal metastatic tumor deposits (Fig. 3C). In cases such as this in which ventricular compliance is poor, we propose that the relative ease with which suture closure occurs is due to a relative propensity to develop an accumulation of CSF in the subarachnoid spaces over the surface of the brain. In infants, whose fontanels and sutures are patent, early widening of the sutures would be expected, resulting in a low-pressure accumulation of CSF as seen in benign external hydrocephalus. Conversely, in adults and older children in whom the sutures are fused or are tightly apposed, a pseudotumor cerebri–like picture with high pressure and no enlargement of the ventricles or subarachnoid space would occur. Pseudotumor cerebri is exceptionally rare in children younger than 3 years of age.13 and benign external hydrocephalus is usually self-limiting by 18 months of age.14 It has been proposed that in both conditions CSF absorption into the venous sinuses is impaired but that in infants the presence of open cranial sutures may allow a transient non-hydrostatic loading of brain parenchyma, resulting in mild nonprogressive macrocephaly rather than pseudotumor cerebri.13 We suggest that our intermediate scenario may arise in young children with reduced ventricular compliance, typically those between 3 and 6 years of age, in whom raised ICP and external hydrocephalus coexist due to the high ICP required to cause sutural diastasis.

This case report should alert clinicians to the potential for early development of raised ICP in children with diffuse pontine glioma. Younger children may present with minimal or no symptoms of raised ICP and may have only mild ventricular enlargement with elevated CSF pressure, which may affect the prognosis of children with loss of vision may have only mild quality of life. Edema is potentially avoided by early recognition and by performing intervention such as treatment.}


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ventricular enlargement on MR imaging, despite grossly elevated CSF pressure that may threaten vision. Although the prognosis of diffuse pontine glioma is extremely poor, loss of vision may have a profound impact on the patient’s quality of life. Early recognition and treatment of this potentially avoidable complication is desirable.

References


Manuscript received March 23, 2004.
Accepted in final form August 31, 2004.
Sources of Funding: RJE is supported by the Mansell Research Fellowship of the Royal College of Surgeons of England. Address reprint requests to: Ian K. Pople M.D., Department of Neurosurgery, Frenchay Hospital, Frenchay Park Road, Bristol BS16 1LE, United Kingdom. email: i.k.pople@hotmail.com.