Elevated intracranial pressure from cerebral venous obstruction by Masson’s vegetant intravascular hemangioendothelioma

Case report

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The case is described of a 15-year-old girl with papilledema and visual obscurations caused by a rare lesion, Masson’s vegetant intravascular hemangioendothelioma, within the venous sinus at the torcular herophili. This lesion impeded cranial venous outflow, leading to intracranial hypertension.

KEY WORDS • hemangioendothelioma • pseudotumor cerebri • intracranial pressure • optic nerve fenestration

RECENTLY, we encountered an unusual case of intracranial hypertension arising from impaired cerebral venous outflow as a result of a lesion at the torcular herophili. The lesion was shown pathologically to be Masson’s vegetant intravascular hemangioendothelioma, a rare pathological entity usually associated with a benign course. This patient presented a major management problem in view of progressive visual deterioration associated with a lesion of uncertain nature at an unusual location. The successful management of this patient by neuro-ophthalmological and neurosurgical intervention is described along with a discussion of the pertinent pathophysiology of the lesion.

Case Report

This 15-year-old girl presented to her local physician with a 6-month history of pressure in her eyes and at the back of her head. She had been diagnosed with paranoid schizophrenia 12 months earlier and was being treated with perphenazine and benztropine. Her medical history was significant for chronic otitis media as a child. She had no history of exposure to vitamin A, retinoids, tetracycline, or other agents that have been associated with pseudotumor cerebri. One month prior to her presentation she noticed a small scotoma in her left eye, and her local physician observed marked papilledema.

Examination. The patient was referred for neuro-ophthalmological evaluation. She reported occasional transient obscurations of vision. Visual acuity was 20/20 in each eye. Contrast sensitivity testing was abnormal bilaterally, worse on the right than the left. Funduscopic examination showed optic nerve-head elevation of 5 diopters on the right and 3 diopters on the left. Visual fields showed bilateral enlargement of the physiological blind spot.

An orbital computerized tomography (CT) scan was obtained and showed borderline dilatation of the optic nerve sheaths but was otherwise unremarkable. A magnetic resonance (MR) image showed a small 2 × 1.5-cm enhancing lesion at the torcular herophili compatible with a dermoid cyst or meningioma. There was no evidence of hydrocephalus, and signal characteristics of the blood proximal to the lesion in the sinus indicated slowing of blood flow rather than complete obstruction as in venous thrombosis. A lumbar puncture showed an opening pressure of 37 cm H2O, and cerebrospinal
fluid analysis revealed 40 mg/dl protein and a normal cell count. A digital subtraction angiogram disclosed narrowing at the confluence of the sagittal and transverse sinuses. The mass at the torcular was of insufficient size to be capable of producing direct elevation of intracranial pressure (ICP) to the degree seen; however, it was deemed enough to compromise venous outflow from the cerebral venous sinuses and to cause the symptoms.

Initially, the patient was followed closely and her neuroleptic medications were stopped without any obvious effect on her papilledema. Her papilledema did reduce slightly over the next few months but did not resolve, and reinstitution of her neuroleptic drugs caused no change in the optic nerve-head elevation. Repeat MR imaging 8 months after presentation showed definite enlargement of both optic nerve sheaths and an increase in size of the torcular mass (Fig. 1). Visual fields at this time showed increasing enlargement of the blind spot approaching fixation, especially on the left. The papilledema had increased but no hemorrhages were found. There was a slight sixth nerve paresis, mild dysmetria and intention tremor bilaterally, and clumsy heel-shin and tandem gait. Transfemoral cerebral angiography showed slow flow through the superior sagittal sinus. There was a capping defect suggestive of an intrasinus lesion and a dense tumor blush arising from a large posterior branch of a middle meningeal artery. It was considered that the lesion most likely represented a meningioma.

Operations. In view of the patient’s progressive visual deterioration and the uncertainty whether resection of the torcular lesion would relieve (and might, in fact, exacerbate) her ICP, we performed bilateral optic nerve-sheath fenestrations via a medial orbital approach prior to intracranial surgery. One week postoperatively the papilledema was notably improved, at which point she underwent an occipital and suboccipital craniotomy. At the time of surgery, a $3 \times 2 \times 1$-cm purplish-red mass within the lowermost portion of the superior sagittal sinus was identified and debulked. A subdural monitor was placed to allow postoperative ICP measurements in case thrombosis of the major venous sinuses resulted from this surgical intervention. A long-term ICP monitoring system was also implanted. It was not believed that complete resection of the lesion should be undertaken, as frozen-section pathological examination of the lesion indicated a benign process and complete resection would necessitate some sinus reconstruction or bypass which carried a high likelihood of postoperative thrombosis.

Postoperative Course. The patient made an uneventful recovery. Initial ICP measurements ranged from 20 to 30 cm H$_2$O. A postoperative MR image showed increased venous flow through the torcular. Three months postoperatively, her headaches and visual symptoms had resolved and neurological examination was normal except for some slight blurring of the optic nerve-head margins and slight elevation on the right. At this time the implanted monitor revealed an ICP of 21 cm H$_2$O. Repeat MR imaging 6 months after surgery showed no recurrence. Eleven months postoperatively, the patient remains asymptomatic and the ICP monitor readings are 10 cm H$_2$O.

Pathological Examination. Histopathological study of the lesion showed multiple sinus lumina filled with angiomatous tissue having racemose thin-walled vascular channels of varying sizes. Several sinus lumina

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\* ICP monitoring system manufactured by Radionics, Inc., Burlington, Massachusetts.
Masson’s vegetant hemangioendothelioma

contained recent thrombi and one of them was associated with papillary endothelialized vegetation (Fig. 2). The histological features of benign angiomatic tissue filling the sinus lumina were compatible with Masson’s vegetant intravascular hemangioendothelioma.

Discussion

Pathological Findings

Masson’s vegetant intravascular hemangioendothelioma is a rare lesion and only a handful of intracranial cases have been reported.4,10,12,20 This entity was first described in 1923 by Pierre Masson,14 in an inflamed hemorrhoidal plexus. Usually considered to be a benign lesion, it has been described in numerous tissues, especially the skin, naso-oral pharynx, larynx, gastrointestinal tract, liver, and pelvic structures. There has been a female preponderance in the cases reported. The pathogenesis of the lesion remains controversial. Masson considered that the lesion was a benign proliferation of endothelial cells with secondary thrombosis and fibrin deposition. Others have suggested that the pathological changes seen were due to an excessive reaction to a normal reorganizational process in a thrombus rather than a neoplastic process.1,3,5,9,18

Masson’s vegetant intravascular hemangioendothelioma may represent part of a spectrum of lesions including intravascular papillary endothelial hyperplasia11,12 or intravenous atypical vascular proliferation.16 However, it must be clearly distinguished from true malignancies such as angiosarcoma, Kaposi’s sarcoma, and angiolymphoid hyperplasia with eosinophilia.13,18 In Masson’s vegetant intravascular hemangioendothelioma, the organizing thrombi develop endothelialized papillary-like projections into the vessel lumen. Unlike angiosarcomas, these structures are confined to the lumen itself. The paucity of mitoses, foci of necrosis, and solid cellular areas without vessel formation also help to distinguish this entity from angiosarcoma.

The only other previously reported case with long-term follow-up monitoring was also from this institution.19 This was a 16-year-old girl with neurocutaneous Masson’s vegetant intravascular hemangioendothelioma and medically intractable seizures with multiple intracerebral, cutaneous, and pulmonary lesions. She was initially believed to have metastatic disease and underwent an excisional biopsy of a cortical lesion, at which time the benign nature of her disease was established. She continued to suffer severe seizures, and 2 years later underwent resection of a mesial temporal lobe Masson’s vegetant intravascular hemangioendothelioma. After 9 years, she remains seizure-free with no evidence of progression of the other intracerebral lesions and no recurrence of her excised lesions on follow-up CT scans.

This previous case and the benign course seen with cutaneous and systemic cases of Masson’s vegetant intravascular hemangioendothelioma suggest that the prognosis for the present patient is good. However, two cases of intracranial Masson’s vegetant intravascular hemangioendothelioma reported in infants showed an aggressive course; a 12-day-old infant suffered a major recurrence 2 months after resection of the lesion,20 and a 14-week-old infant showed progressive enlargement of her lesion resulting in death 6 months later.4 No long-term follow-up data were available in the 55-year-old woman reported by Izukawa, et al.;19 she presented with an intracerebral hematoma associated with a cavernous angioma and intravenous papillary endothelial hyperplasia.

The accurate identification of Masson’s vegetant intravascular hemangioendothelioma and its distinction from malignancies with which it may be confused are stressed. The accurate identification of this lesion in our patient has so far avoided the need for further therapy.

Ophthalmological Aspects

Our patient had in effect a similar problem to pseudotumor cerebri albeit not from an idiopathic source. The major important neurological complication of pseudotumor cerebri is visual loss secondary to papilledema. Optic nerve sheath fenestration or sheathotomy has been used successfully to treat pseudotumor cerebri associated with progressive visual loss.3,6,1,19

The patient’s deteriorating vision along with grave concerns as to possible increased venous obstruction by disease progression or direct surgical intervention led us to carry out optic nerve-sheath fenestration as an initial procedure. By this means her vision was protected in the interim and also prophylactically, should she develop further intracranial hypertension from increased venous obstruction. The improvement in papilledema the week following sheathotomy suggests that, even if adequate venous outflow from the sinuses was not maintained, the optic nerves were at least partially protected. Partial excision of the lesion appears to have improved the venous outflow from the sinuses, as shown on MR imaging, and was sufficient to relieve both her visual and neurological symptoms as well as lower her ICP.

As in cases of pseudotumor cerebri where visual deficits predominate, the papilledema in this patient responded well to optic nerve-sheath fenestration. Our experience here suggests that this maneuver may be helpful in cases of visual loss from papilledema when the primary cause of the papilledema may not be amenable to direct therapy.

Neurosurgical Aspects

Intracranial hypertension secondary to cerebral venous outflow obstruction is a well-recognized entity.2,7 Rosman and Shands12 and Haar and Miller6 have reviewed this subject. They were able to identify two different groups of patients. One group includes infants who appear to develop hydrocephalus especially if there is generalized elevation in the intracranial venous pressure (such as from a superior vena cava syndrome,
frequently related to central venous-line induced thrombosis). The other group includes older children and adults who tend to develop pseudotumor cerebri-like syndrome and in whom normal pressure may still be present in some part of the intracranial venous system (such as from sagittal or lateral sinus thrombosis where alternative drainage pathways via scalp, facial, vertebral, and intracranial collateral venous channels may still be open). Our patient clearly falls into the latter group. Through the ability of our present technology to follow the size of the lesion and to monitor her ICP noninvasively, this patient provided a unique opportunity to allow long-term assessment of cerebral venous obstruction and its effect on ICP and may help shed further light on this unusual phenomenon.

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