

An uncommon cause of increased intracranial pressure

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Abstract

Patients with increased intracranial pressure frequently present to ophthalmologists with a variety of neurologic complaints. This report describes a patient with congenital cerebrovenous malformation who developed increased intracranial pressure following unilateral radical neck dissection. The pathogenesis, diagnostic investigations, interpretation, and subsequent management of this condition are discussed.

Key words: Intracranial hypertension, Neck dissection

Introduction

Patients with increased intracranial pressure (ICP) present to ophthalmologists with a variety of complaints, such as visual disturbances and diplopia from the false localizing sign of sixth nerve palsy. This report describes a patient with an unusual cause of increased ICP following unilateral radical neck dissection.

Case report

A 40-year-old Chinese man received a diagnosis of nasopharyngeal carcinoma in 1993. He was treated with radiotherapy, and subsequently had remission of the disease. Local recurrence occurred in 1996. He was treated with a second dose of radiotherapy, which was complicated by radiation necrosis of both temporal lobes. Long-term low-dose oral steroid (prednisolone 5 mg/d) was given to control the cerebral edema. He presented again in July 1998 complaining of a right submandibular mass. Fine-needle aspiration cytology showed lymph node involvement by

a metastatic undifferentiated carcinoma. The source of the primary tumor could not be found. Right radical neck dissection was performed in the same month, with the removal of the internal and external jugular veins, submandibular lymph nodes, and part of the sternocleidomastoid muscle.

On postoperative day 2, he complained of mild frontal headache, which developed into a generalized headache with nausea 2 weeks later. He also noticed horizontal diplopia at the extreme left gaze at distance. He was referred to the Hong Kong Eye Hospital at this stage. On examination, his corrected Snellen visual acuity was 0.8 in both eyes. His pupils were equal and reactive, and no relative afferent pupillary defects were present. He was able to read only the first plate of the Ishihara chart. Examination of the visual fields showed enlarged blind spots bilaterally. No other field abnormalities were present. Motility examination revealed impaired left abduction, which was consistent with left sixth nerve palsy. All the other cranial nerves were normal, but there was bilateral disc swelling with flame hemorrhages.

As the clinical features were suggestive of raised ICP, a computed tomography (CT) scan was ordered (**Figure 1**). CT showed old bitemporal radiation necrosis without evidence of space-occupying lesions or ventricular dilatation. The nasopharyngeal area was clear. Magnetic resonance imaging (MRI) provided no additional information. Lumbar puncture was performed with an opening pressure of 42 cm of water. The cerebrospinal fluid profile, including protein, cell count, and glucose was normal.

Given the fact that this patient had increased ICP without evidence of a space-occupying lesion and ventricular dilatation, and a normal cerebrospinal fluid profile, pseudotumor cerebri (PTC) was one of the differential diagnoses.¹ However, it is unusual for this condition to present suddenly

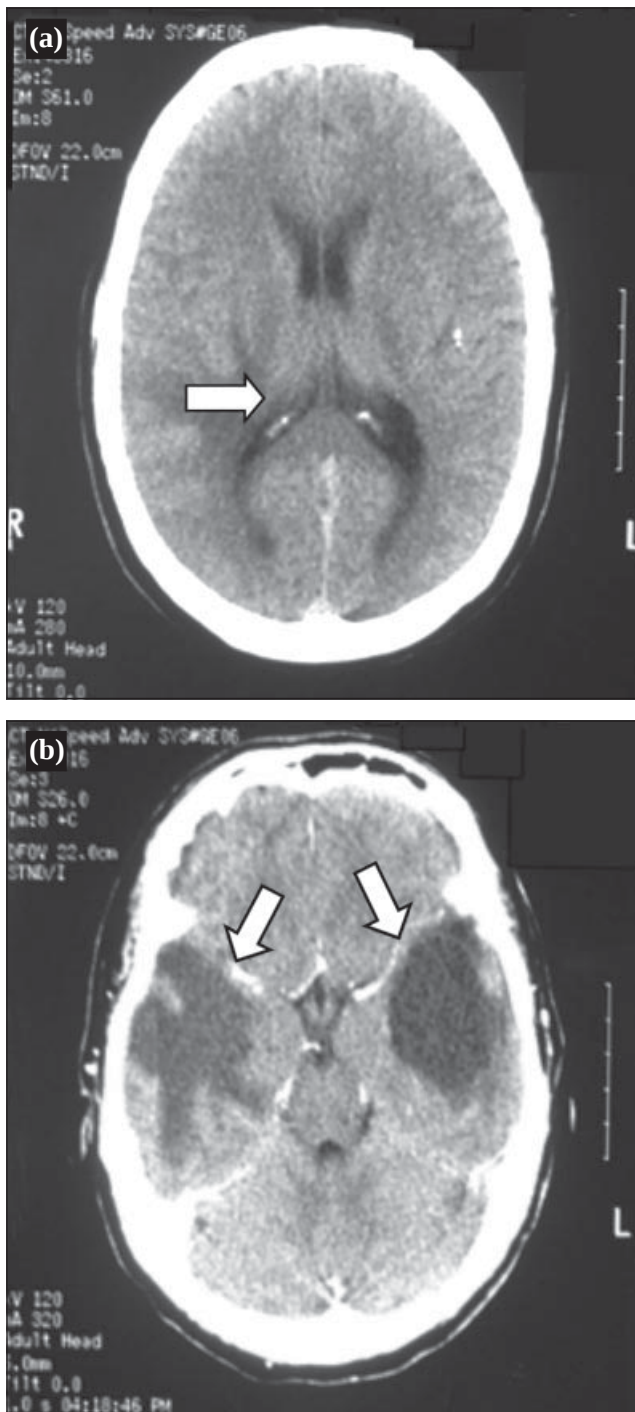


Figure 1. Computed tomography scan of the brain showing (a) normal size ventricles (arrow) and (b) old radiation necrosis of the temporal lobes bilaterally (arrows). No space-occupying lesions were seen.

in a man who had recently undergone a major operation. A rare cause of raised ICP would be venous drainage insufficiencies such as superior sagittal sinus thrombosis that may have a similar clinical presentation.

A magnetic resonance venogram (MRV) was obtained to exclude this possibility (**Figure 2**). The MRV showed obstruction at the origin of the right internal jugular vein (IJV) and hypoplastic left transverse sinus. All other major intracranial venous sinuses appeared patent. Obstruction of the

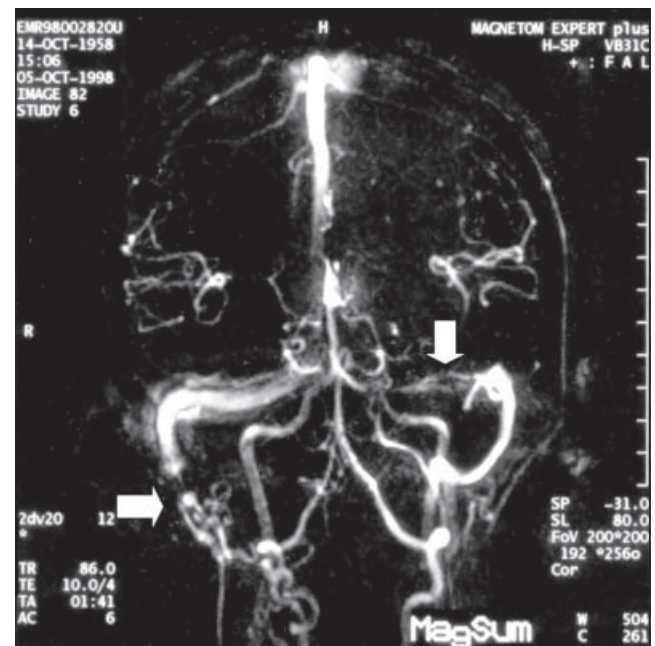


Figure 2. Magnetic resonance venogram showing an obstruction at the origin of the right internal jugular vein (horizontal arrow) and hypoplastic left transverse sinus (vertical arrow). All other major intracranial venous sinuses appeared patent.

right IJV was likely to result from the previous right radical neck dissection with ligation. Regardless of the underlying pathogenesis for the apparently ‘hypoplastic’ left transverse sinus, immediate treatment was needed to lower the ICP with the aim of preserving vision. An oral carbonic anhydrase inhibitor (acetazolamide 250 mg 4 times daily) and a diuretic (frusemide 40 mg daily) were given. The headache and papilloedema persisted, and the ICP remained elevated. A lumbo-peritoneal shunt was eventually inserted in December 1998. The headache subsided following the insertion of the shunt, and all the subsequent ICP measurements were within normal limits. The papilloedema gradually resolved but the patient continued to have residual left sixth nerve palsy.

Discussion

Increased ICP is a documented complication of bilateral radical neck dissection with removal of the jugular veins.² Removal of the IJV necessitates redirection of cerebrovenous blood to collateral pathways. Collateral circulations via the vertebral, pterygoid, and orbital plexus will usually develop, relieving some of the intracranial venous congestion.³ If this collateral system is insufficiently developed, neurologic sequelae due to impaired venous drainage and increased ICP may result.³

Review of the literature showed 2 possible explanations for the hypoplastic left transverse sinus. Bono et al suggested that most people have a dominant right transverse sinus that appears larger than the left transverse sinus on MRV, giving the false impression of a hypoplastic left transverse sinus.⁴ Whether this hypoplastic left transverse sinus is sufficient for cerebrospinal fluid drainage after ligation of the outflow

pathway of the dominant right transverse sinus is unclear. On the other hand, King et al suggested that acquired narrowing of the transverse sinus might be a consequence, rather than a cause, of increased ICP.⁵

This report illustrates the importance of MRV in the identification of the underlying cause in this patient. In the pre-MRV era, patients presenting with increased ICP, normal cerebrospinal fluid profiles, without hydrocephalus and mass lesions, would have a diagnosis of PTC syndrome in accordance with the Modified Dandy's Diagnostic Criteria.¹ However, PTC could be due to multiple causes, including intracranial sinus venous abnormalities/thromboses or idiopathic causes. Clinical differentiation between intracranial sinus-related intracranial hypertension and idiopathic intracranial hypertension is difficult. Sylaja et al compared 2 groups of patients — 1 group with MRI or magnetic resonance angiography (MRA)-confirmed intracranial sinus venous thrombosis, and the other in whom venous thromboses had been excluded.⁶ The clinical findings were the

same for the 2 groups except for female sex and lower cerebrospinal fluid protein level, which were significantly associated with the latter group. MRA should therefore be performed to exclude other causes before diagnosis for these patients with idiopathic disease.⁶

Although isolated cases of raised ICP following unilateral radical neck dissection have been reported,^{2,3,7-9} only 1 had neuroimaging to document the neurovascular malformations.² The MRV of the patient reported here provides independent documentation of the venous anomaly.

The optimal management for raised ICP for this patient remains unknown. The usual steps of diuretics and lumbar puncture were insufficient for control of ICP in this patient. A shunt was eventually needed. One might have postulated that other surgical interventions such as stenting,¹⁰ lumboperitoneal shunting, or optic nerve sheath fenestration may also be beneficial. Further research will help to answer this question.

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