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Case Report

Intracranial Hypertension Secondary to Cervical Dural Arteriovenous Fistula

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Abstract

Idiopathic intracranial hypertension (IIH) is a disease of mainly unknown etiology. Latest theories as to the pathogenesis have postulated a final common pathway of cerebral venous hypertension secondary to venous outflow impairment leading to decreased cerebrospinal fluid absorption. We present the case of a 42-year-old female who was treated for several years for headache and for approximately 12 months for IIH until appropriate imaging showed a right-sided cervical dural arteriovenous fistula (AVF) at the level of C4. The patient's IIH symptoms resolved following surgical excision of the fistula. We suggest that the cranial venous outflow impairment secondary to the cervical AVF was responsible for intracranial hypertension and that complete investigation of IIH patients should include imaging of the neck vasculature.

Keywords: *Arteriovenous fistula, arteriovenous malformation, benign intracranial hypertension, Idiopathic intracranial hypertension, pseudotumor cerebri*

Introduction

Idiopathic intracranial hypertension (IIH) is a disease of unknown etiology usually affecting young females. It is characterized by headaches, visual disturbances, and papilledema and has a variable course.[1,2] In the most severe cases, it can lead to significant and/or permanent loss of visual function.

Several types of pathogenesis have been implicated in IIH. While it was previously thought that disturbed cerebrospinal fluid (CSF) dynamics were responsible, recently increased cerebral venous hypertension secondary to venous outflow impairment leading to decreased CSF absorption has also been implicated as a potential common final pathway in the pathophysiology of IIH.[3] It is, therefore, conceivable that other venous abnormalities resulting in reduced venous outflow can cause intracranial hypertension. We present a case of symptomatic intracranial hypertension secondary to a cervical arteriovenous fistula (AVF) which settled with surgical treatment of the AVF.

Case Report

In 2013, a 42-year-old female teacher with a body mass index of >30 was referred to the neurosurgical clinic by her neurologist, who was seeing her for symptoms related to IIH, to consider CSF diversion for medically refractive visual symptoms. The patient had bilateral papilledema and was complaining of frontal headache behind both eyes, sensitive to coughing, persistent nausea and vomiting, general lethargy, imbalance, poor concentration, and visual blurring. On examination, hemorrhage, hydrocephalus, space-occupying lesions, and venous sinus thrombosis were ruled out by imaging of the brain. Her neurological examination was normal with regard to limbs and cranial nerves. Assessment of the optic system showed normal color vision and visual acuity but infranasal field loss on the right and bilateral papilledema. A lumbar puncture (LP) was performed with an opening pressure was 33 mm H_2O, and her symptoms improved significantly.

In her past medical history, the patient had been suffering from headaches since her first pregnancy in 2000 and since 2006 had been managed by a neurologist. In 2007, she had already been referred to a neurosurgeon since a left posterior frontal arachnoid cyst had been found on imaging of the brain. Although it was thought at the time that the cyst would be unlikely to be the cause of her headaches, a burr hole marsupialization was undertaken in 2008 which resulted in several months of good pain relief. Following on from this,

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the patient underwent a mini-craniotomy with further marsupialization of the cyst and obtained good symptom relief for further 6 months. Then, however, the symptoms reoccurred over the course of the next 2 years. With the cyst having been treated, the diagnosis of cluster headaches (responding well- to high-flow oxygen therapy) was made, and the patient attended a residential pain management programme in 2010 with good response.

Furthermore, in 2012, the patient was involved in a camping gas canister explosion during which she was thrown 8 feet, landed on her back, and hit her head. Although she recovered well from her minor external injuries, her neurological problems in the way of headaches, nausea and persistent vomiting, and dizziness were exacerbated again. LPs repeatedly showed increased opening pressure. Finally, a diagnosis of IIH was made and acetazolamide commenced, but the patient developed poor concentration and a skin rash and acetazolamide had to be stopped. Even with topiramate and furosemide, the symptoms persisted and control was only possible with repeated LPs, all of which showed increased opening pressure. Eventually, the patient was referred back to neurosurgery for consideration of CSF diversion, where she presented with the signs and symptoms described above.

Further imaging was performed. Computed tomography (CT) venogram revealed a high-grade short-segmented stenosis at the right sinusoidal knee, a longer segmented stenosis of the distal left transverse sinus, and significant narrowing of the left sigmoid sinus close to the jugular foramen. Formal digital subtraction angiogram (DSA) and further superselective spinal angiography showed dorsal medullary early venous shunting in the form of a right-sided AVF at the level of C4 fed by the radiculomeningeal artery at C4/5 [Figure 1].

The dorsal medullary vein drained into the right transverse sinus, likely causing venous hypertension [Figure 2]. Due to narrow vessel diameter, it was not possible to embolize this AVF endovascularly. The fistula was surgically divided in 2013 by means of a cervical C4/5 laminectomy and excision of communicating blood vessels. Initially, this resulted in some resolution of the symptoms with the improvement of the field defect and complete disappearance of the papilledema.

Postoperatively, the patient's headache, nausea and vomiting improved considerably as did the visual fields.

On 2-year follow-up, the patient's neurological improvement is maintained.

Discussion

IIH is a disease of unknown etiology usually affecting young females of reproductive age. The annual incidence is higher in obese women ranging from 4 to 21 per 100,000 in the age group of 15–44.[4] IIH has a variable course: it is either self-limiting over a few years or has a protracted course over many years.[2] The disease features are headaches, visual problems, and papilledema with severe or permanent loss of visual function in up to 25% of the cases.[5] Occasionally, patients present with other, nonspecific symptoms such as tinnitus, dizziness, nausea, and lethargy sometimes making IIH difficult to distinguish from other forms of headaches such as migraine or cluster headaches. The diagnosis is made with the help of clinical examination (evidence of papilledema) and high opening pressure on LP with resolution of symptoms on reducing the CSF pressure during the puncture.

The underlying pathophysiology is so far unknown, but several different theories have been put forward, mostly based on disturbed CSF dynamics. Whether the condition is due to increased CSF production at the choroid plexus as suggested by Quincke[6] or restricted CSF drainage at

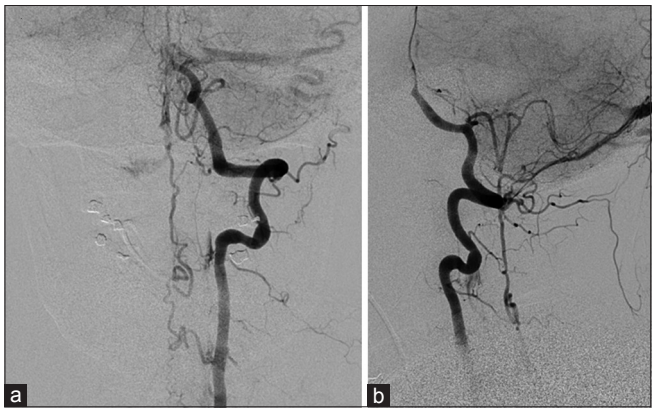

Figure 1: (a and b) Lateral and anteroposterior digital subtraction catheter angiogram of the left vertebral artery demonstrating the fistula (white arrow) with dorsal medullary venous drainage (white arrowhead) and early dural sinus venous filling (black arrowhead)

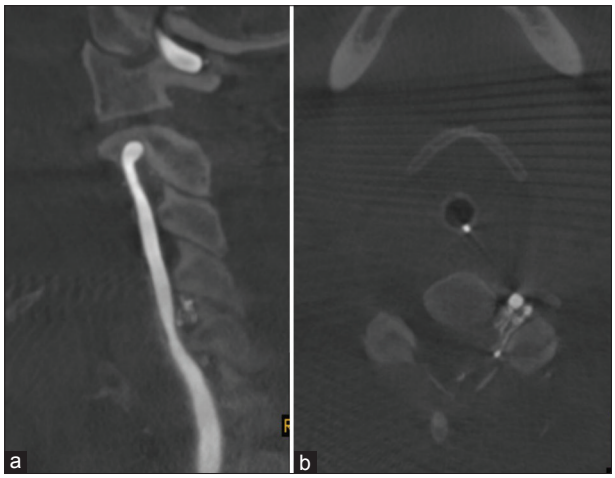

Figure 2: (a and b) Lateral and anteroposterior reformats of digital subtraction of the left vertebral artery demonstrating fistulous point at the C4 exit root foramen on the dural surface. The site of fistula with radiculomeningeal feeders (white arrow) and the filling of the dorsal medullary vein (arrow head)

the arachnoid villi is the subject of much speculation.[1] According to the Monro-Kellie-Doctrine, any increase in the volume of blood, CSF or brain, or anything impeding CSF or venous outflow would increase ICP. Dandy[7] proposed that an increase in cerebral blood volume could be responsible for raised intracranial pressure. Others studies found an increase in brain cell water content in patients with IIH,[8] and more recently, in white matter water signals.[9] A further theory is obstruction of CSF or venous outflow as a cause of raised intracranial pressure. The underlying mechanism suggested is that increased sagittal sinus pressure results in decreased CSF absorption as higher CSF pressure is necessary to drive bulk flow of CSF across the meninges.[10] Cerebral venous hypertension secondary to venous outflow impairment causing reduced CSF reabsorption has, therefore, been implicated as a potential common final pathway in the pathophysiology of IIH.[3]

Following this hypothesis, evidence of good treatment results in IIH with the help of venoplasty or venous stenting has been reported in the literature. These interventions may be alternatives to CSF diversion by means of a CSF shunt.[11]

Secondary intracranial hypertension has been described as a result of venous sinus thrombosis or mass lesion, for example, meningioma causing venous sinus compression.[12] A case series of dural AVF causing intracranial hypertension has been reported.[13]

Taking the above into consideration, we speculate that it is possible that extracranial vascular malformations resulting in impaired venous outflow from the brain can cause intracranial hypertension. We present a case of intracranial hypertension secondary to a cervical AVF which settled with the treatment of the AVF. We believe, however, that this is the first case report of intracranial hypertension caused by a vascular abnormality outside of the cranium and treated with removing of the abnormality.

Our patient is clearly complex in that she had a pre-existing diagnosis of cluster headaches and a history of posttraumatic exacerbation of her symptoms. However, the usual diagnostic modified Dandy criteria for IIH, i.e., visual impairment with papilledema, headaches, raised intracranial pressure on LP, and subsequent resolution of symptoms, were being met. Indeed, the patient had moderate treatment success with Acetazolamide but could not tolerate the side effects. Repeated LPs controlled the symptoms for a period of time.

The definitive treatment of the cervical AVF resulted in satisfactory continued resolution of the IIH symptoms. This lends support to the hypothesis of increased venous sinus pressure secondary to obstructed cranial venous outflow.

An interesting point is whether the gas explosion the patient was involved in was contributory to the symptoms. This accident could have resulted in deep tissue injury restricting the venous outflow form the brain.

In our institution, patients suspected of having IIH have at least a CT angiogram (4D), but more often a formal DSA with assessment of the venous phase. During this, the caliber of the cranial venous sinuses can be measured as well as the pressure across any stenosed segments of sinus. Furthermore, venous sinus venoplasty is possible.

However, due to the known association of IIH with vascular malformations and the success in the treatment of our patient, we think that formal angiography of the intracranial vessels as well as the neck should be included in the standard workup of IIH patients during the ascertaining of the diagnosis.

Conclusion

Cranial venous outflow impairment, in this case secondary to the cervical AVF, can be responsible for intracranial hypertension. Treatment of the outflow impairment resolves intracranial hypertension, and therefore complete investigation of IIH patients should include imaging of the neck vasculature.

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Conflicts of interest

There are no conflicts of interest.

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