

Balloon angioplasty for venous sinus stenosis in a idiopathic intracranial hypertension case

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Abstract

The Idiopathic Intracranial Hypertension (IIH) is a well characterised condition with intractable headaches, visual obscurations, and papilloedema as dominant features, mainly affecting obese women. With the advent of magnetic resonance (MR) venography and increased use of cerebral angiography, there has been recent emphasis on the significant number of patients with IIH found to have associated non-thrombotic dural venous sinus stenosis. This has led to a renewed interest in endovascular stenting and angioplasty as a treatment for IIH in patients non-responsive to medical treatment. We present a patient without known risk factors for IIH and non-responsive to treatment. The 19-year-old woman presented with headache and diplopia. She was diagnosed with IIH since she was five years of age and had been non-responsive to lumbar cerebrospinal fluid (CSF) drainage and acetazolamide treatment. MR venography revealed thin calibration of transverse sinus. Digital subtraction angiography (DSA) venous phase also revealed 50% stenosis of transverse sinus, 50% stenosis of left proximal sigmoid sinus and 90% stenosis of its distal part leading to obstruction of left transverse sinus outflow and forced directed drainage of left hemisphere to the anterior region.

Keywords: Idiopathic intracranial hypertension, Venous sinus stenosis, Endovascular therapy.

Introduction

The syndrome of increased intracranial pressure without hydrocephalus or a mass lesion and with normal cerebrospinal fluid (CSF) findings, previously referred to as pseudotumour cerebri (PTC) or benign intracranial hypertension, is a diagnosis of exclusion now termed idiopathic intracranial hypertension (IIH).¹ Quincke first described the disorder "meningitis serosa" in 1893 in patients who had increased intracranial pressure without a brain tumour. More commonly, symptoms of IIH that reflect generalised intracranial hypertension include

headache, feeling of pulsatile intracranial sounds, and diplopia and, less frequently, neck pain, back and shoulder pain, or radicular pain. Other non-specific symptoms of meningeal irritation may be present, including photophobia, nausea and vomiting.^{1,2} With the development of neuroimaging techniques like digital subtraction angiography (DSA) and magnetic resonance imaging (MRI) venography, IIH related to venous system anomalies are detected more frequently. Recently, in addition to the arterial system, the venous system also is being a target for endovascular treatment options. Dilatation of one of the sinuses with a stent or balloon angioplasty treatment reduced the pressure gradient, with striking symptomatic improvement in patients that do not respond to medical treatment.³

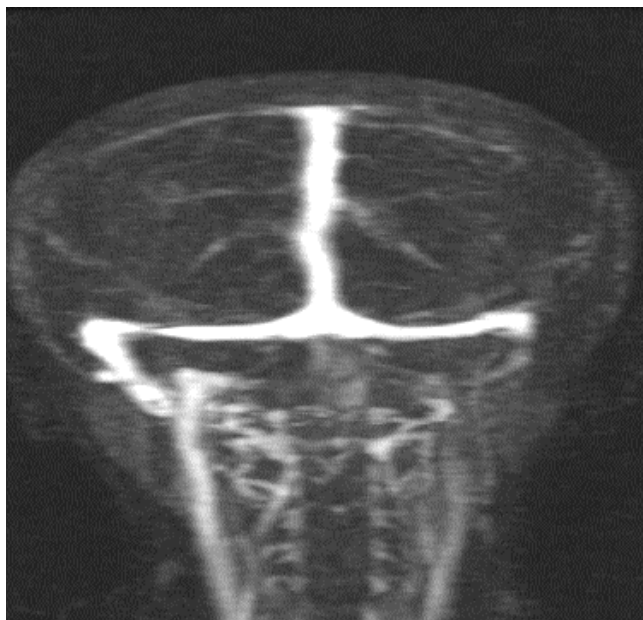
We present a case to show the benign outcome in a patient with IIH who was non-responsive to medical treatment, and who underwent balloon angioplasty recanalisation in the right transvers sinus and left sigmoid venous sinus.

Case Report

A 19-year-old woman presented with headache and diplopia. She was diagnosed with idiopathic intracranial pressure since 5-year-old and had been non-responsive to lumbar CSF drainage or acetazolamide treatment. The physical examination was normal with normal body mass index (BMI). Neurological examination revealed normal findings, but bilateral optic atrophy. In order to exclude possible different aetiologies we also tested for markers of vasculitis, and the results were negative. Opening pressure of lumbar CSF was 250mm/H₂O. The CSF examination showed mild pleocytosis (lymphocytes 10/mm³), elevated protein levels (56.8 mg/dL) and negative cultures. The CSF Ig-G index was within normal limits and negative CSF oligoclonal banding. Viral panel (Ebstein-Barr virus, Varicella zoster virus, Cytomegalovirus, herpes simplex virus type 1, 2) were negative. Finding of cranial MRI were normal. MRI venography revealed thin calibration of transverse sinus (Figure-1). DSA venous phase also revealed 50% stenosis of transverse sinus, 50% stenosis of left proximal sigmoid sinus, and 90% stenosis of its distal part leading to obstruction of left transverse

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MRI: Magnetic Resonance Imaging

Figure-1: Pre-treatment MRI Venography.

DSA: Digital subtraction angiography.

Figure-2: Post-treatment DSA venous phase.

sinus outflow and forced directed drainage of left hemisphere to the anterior region. Venography was performed by selective contrast injections in the transverse sinus through the micro-catheter to confirm

the presence and location of stenosis. Balloon catheter dilatation of the left sigmoid sinus was performed. On observing higher flow pattern we terminated our intervention (Figure-2). In order to prevent restenosis the patient's administration with acetazolamide was maintained as presumably elevation of CSF pressure would have led to restenosis. The 8th-12th week follow-up MRI venography did not show restenosis and her ophthalmological assessment showed no progression. Following the procedure except for a headache lateralised to the treated side which settled over days, no other clinical symptom or sign appeared.

Discussion

By definition, in secondary intracranial hypertension there is an underlying medical condition, whereas in IIH, the cause is not known. PTC classically presents with pressure like throbbing, and usually unremitting with retro-ocular pain headache (90%-94%), vision loss with the typical impairment presenting as tunnel vision 68%-85%, pulse-synchronous tinnitus (58%) and other less common symptoms, including photopsia (54%) and orbital pain (44%).⁴ Among the aetiological factors associated with IIH, lateral or superior longitudinal sinus thrombosis, menstrual irregularities with obesity, obstructive sleep apnoea, endocrine disorders, including adrenal insufficiency, hyper-adrenalism, and corticosteroid hormone withdrawal, hyperparathyroidism, hypothyroidism, pregnancy, menarche, lupus, intoxication with vitamin A or chlordecone, tetracycline therapy, spinal cord tumours, and the Guillain-Barré syndrome (GBS) are the most known factors.^{4,5}

In some cases no underline cause can be found. Still in these cases with the development of radiological techniques various pathogenic mechanisms have been considered, including idiopathic intracranial venous hypertension caused by sinus venous stenosis. Many authors propose that any patient with suspected elevated intracranial hypertension undergo MRI venography in addition to traditional orbital MRI to evaluate venous thrombosis or stenosis as the aetiology of PTC symptoms.^{6,7} Recent imaging methods have enhanced detection of intracranial sinus venous stenosis previously undetected due to artifactual flow voids in the transverse sinuses. A study identified venous stenosis in 90% patients with IIH using a novel MRI venography method, auto-triggered elliptic centric-ordered imaging with sensitivity and specificity of 93%.⁷ Our patient being young in age with no known risk factors and non-responsive to previous treatments led us to perform further radiological procedures.

The primary goal of management in PTC is restoration of

visual acuity and resolution of papilloedema. Weight-loss is thought to address one of the fundamental risk factors for recurrence of PTC and it may reduce the risk of PTC and concomitant vision impairment through reductions of central venous pressure or a tentative endocrinological mechanism.⁸

Medications to treat or prevent recurrence of PTC typically have activity against carbonic anhydrase, acetazolamide being historically the most commonly used medication to treat PTC, but topiramate has been shown to have equal effect as a result of partial carbonic anhydrase-inhibition activity augmented by beneficial analgesic action against headaches and weight-loss as a fortuitous side effect.^{4,9} Corticosteroids may result in temporary remission of PTC, but the risks of chronic steroid use may result in hypothalamic-pituitary-adrenal axis suppression, growth suppression, decreased bone mineralisation, cataract formation, aseptic bone necrosis and weight-gain firstly in already obese patients preclude their use.⁹ Our patient's long history of acetazolamide treatment was insufficient in preventing symptoms and occurring of optic atrophy.

Indications for surgical treatment such as CSF shunt insertion, optic nerve sheath fenestration, or subtemporal decompression include medical treatment failure or non-compliance, new or worsening visual field deficits, intractable headache, or fulminant IIH. Shunts have complications, including shunt migration and dislocation, infection, acquired Chiari malformation, and intracerebral haemorrhage. Up to 64% of ventri-culo and lumbo-peritoneal shunts fail within 6 months; re-operation is common for recurrence of papilloedema and high CSF pressures. Optic nerve sheath fenestration has an up to 40% complication rate, including visual loss, motility and pupillary dysfunction, and vascular complications.¹⁰ Such poor results have been tolerated due to a lack of a viable alternative. For medically refractory IIH patients, particularly those with visual loss and intractable headache, surgical options may be considered but, having significant limitations originating from the probable complications has led to think different treatment alternatives.

None of the above surgical treatment was applied to our patients, but, regarding her age in the first place, endoluminal treatment was preferred as an easier and less complicated procedure confirming sinus pathology by MRI venography.

In IIH patients similar to ours whose venous disease was confirmed by radiological techniques and who are unresponsive to medical treatment, the next step is identifying whether the sinus stenosis is the cause or the

consequence of the increased intracranial pressure. In a study, dilatation of stenosis with a stent provided a reduction in intracranial venous sinus pressure which was accompanied by an immediate and sustained clinical improvement, evidence of a causal relation between venous sinus disease and IIH.¹¹ If venous outflow obstruction was the cause, then dilating the stenosis and abolishing the pressure gradient should be curative. If sinus stenosis were secondary to raised intracranial pressure then, in patients in whom stenting was beneficial, these stenosis must have been responsible for a sufficient increase in the intracranial pressure to render them symptomatic. So, regardless of the cause, consequence relationship between the stenosis and the patients symptoms, this procedure being safe and effective alternative to CSF shunt surgery, should be considered.¹²

Our patient was assessed at the 8th-12th weeks, with a follow-up venography. After stenting, except for a headache lateralised to the treated side, which settled over days to weeks, no other clinical symptom or sign appeared. Ophthalmological assessment showed no progression. Acetazolamide treatment was maintained in order to diminish the CSF pressure as the main cause of restenosis.

To date, approximately 40 patients with IIH treated with sinuovenous stent placement or angioplasty have been reported. We aimed not only at demonstrating the similar outcome in our patient, but to emphasise the application of this technique mostly in young, not obese patients and not responding to medical treatment, before taking into consideration of other surgical treatments with higher complication rate. In sinus venous balloon angioplasty treatment, restenosis being the most frequent complication, we also suggest acetazolamide treatment as a maintenance therapy since high CSF pressure may result in restenosis.

Conclusion

The importance of venous sinus disease in the aetiology of IIH is probably underestimated. Patients with IIH in whom a venous sinus stenosis is demonstrated by a non-invasive radiological workup should be evaluated with direct retrograde cerebral venography and DSA. In patients with venous sinuses lesion who experienced medical treatment failure, endovascular treatment can be considered as the treatment of choice due to its lower complication rate.

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