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Case report

Ventriculoperitoneal shunt treatment in a pregnant renal transplant recipient with idiopathic intracranial hypertension: Case report and review of the literature

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ABSTRACT

Idiopathic intracranial hypertension (IIH) is a relatively uncommon disorder characterised by raised intracranial pressure without an established pathogenesis. Diagnosis of IIH requires the demonstration of symptoms and signs referable only to elevated intracranial pressure; cerebrospinal fluid (CSF) opening pressure >25 cm H_2O measured in the lateral decubitus position; normal CSF composition; and no evidence for an underlying structural cause demonstrated by using MRI or contrast-enhanced CT scan for typical patients and MRI and MR venography for atypical patients such as man, children and those with low body mass index. We present a 38-year old primigravid renal transplant patient at 7 weeks of gestation who presented with 2 weeks of intense, throbbing, holocranial headache, nausea, vomiting, photophobia, diplopia and progressive visual loss. When medical treatment fails and/or not appropriate to use due to the reported of teratogenic risks in pregnant women, surgical interventions gain importance. In this particular patient, venticuloperitoneal shunt was chosen as the CSF diversion technique. In this case report indications, contraindications in addition to outcomes regarding headache, vision loss and the resolution of papilloedema of the present surgery options for IIH are discussed.

1. Introduction

Idiopathic intracranial hypertension (IIH) is a relatively uncommon disorder characterised by raised intracranial pressure without an established pathogenesis. Diagnosis of IIH requires the demonstration of (1) symptoms and signs referable only to elevated intracranial pressure; (2) cerebrospinal fluid (CSF) opening pressure >25 cm H_2O measured in the lateral decubitus position; (3) normal CSF composition; and (4) no evidence for an underlying structural cause demonstrated

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by using MRI or contrast-enhanced CT scan for typical patients and MRI and MR venography for atypical patients such as man, children and those with low body mass index (BMI) [1].

Headache is the most prominent feature of the disease. Patients describe different patterns of headaches; pressure-like, holocranial, frontal or retro-orbital which typically worsen with Valsalva-type manoeuvres. Posture-related unilateral or bilateral transient visual obscurations are usually reported. Pulsatile tinnitus, nausea, vomiting, photophobia and diplopia are amongst the other most encountered complaints [2].

We present a pregnant renal transplant recipient which complicated with IIH at the seventh week of gestation.

2. Case report

A 38-year old primigravid woman at 7 weeks of gestation presented with 2 weeks of intense, throbbing, holocranial headache, nausea, vomiting, photophobia, diplopia, progressive visual loss and transient total visual obscurations precipated by changes in posture that last several seconds. Her past medical history was remarkable for an end-stage kidney disease in the course of reflux nephropathy which progressed to kidney transplant from a cadaveric donor nine years ago. The immunsupressive treatment included tacrolimus, azathioprine and prednisone. The allograft function and evolution were good (creatinine = 1.1 mg/dL). On examination she was normotensive and her BMI was 25 kg/m^2. Neurologic examination revealed left abducens nerve palsy, bilateral papilledema with retinal haemorrhages.

In view of the possibility of an intracranial mass lesion, cranial MRI and MR venography were performed which evidenced no structural alterations (Fig. 1a and b). Ophthalmologic examination revealed a visual acuity of 20/20 with normal anterior segment findings of both eyes. The central 30-2 visual field test demonstrated an enlarged blind spot and retinal haemorrhages with optic disc swelling in both eyes. A diagnostic lumbar puncture (LP) was performed. The opening pressure which was recorded with a simple column manometer was 340 mm H_2O. Cultures and investigations for toxoplasmosis, tuberculosis, syphilis, neurocysticercosis, cryptococcosis, and cytomegalovirus in the CSF were also planned in case of a central infection associated with chronic immunsuppression were all negative. Acetolazamide could not be initiated due to slightly elevated serum creatinine levels and ongoing pregnancy. She underwent repeated lumbar punctures, however CSF pressure continued to increase and her vision deteriorated. With the concern of ongoing visual loss besides intractable headache, the patient was consulted with the neurosurgery department. A ventriculoperitoneal (VP) shunt was placed on the 14th day of her referral. She experienced instant relief, remained asymptomatic and was discharged the following day. Pre- and postoperative photographs of the fundus are illustrated (Fig. 2a and b).

Unfortunately on the course of this process the patient aborted at the tenth week of gestation spontaneously after the insertion of VP shunt. The healing process accelerated significantly after the miscarriage. Considering the headache and vision, she is still asymptomatic after 3 years of the diagnosis of IIH.

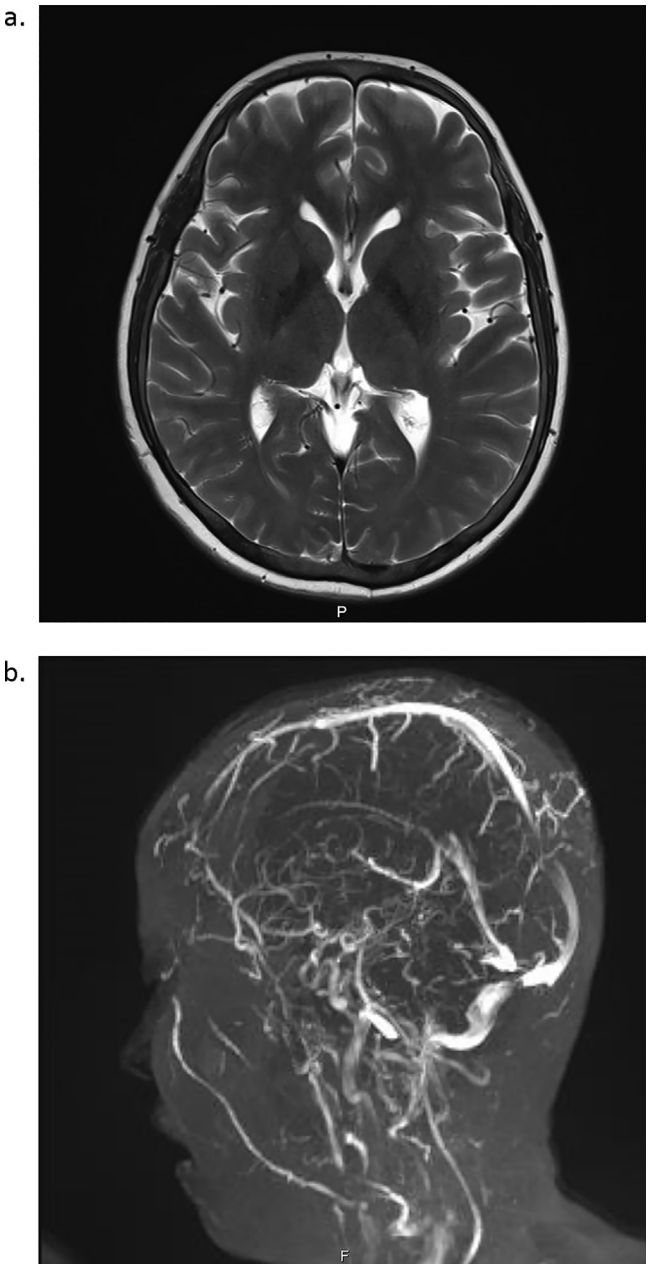

Fig. 1 – (a) Cranial MRI. (b) MR venography.

3. Discussion

The incidence of IIH in renal transplant patients is unknown and the pathogenesis still remains unclear [1,3]. Proposed mechanisms are parenchymal oedema, increased cerebral blood volume, excessive CSF production, venous outflow obstruction and compromised CSF resorption. Possible contribution of inflammatory factors is also being discussed in very recent studies. One of these is a study in which cytokine levels and oligoclonal bands have been found to be correlated with IIH and loss of vision. Results of this prospectively designed study emphasizes the immunologic background of IIH [4].

Previously, it was believed that IIH was triggered or exacerbated by pregnancy. However, IIH occurs in pregnancies

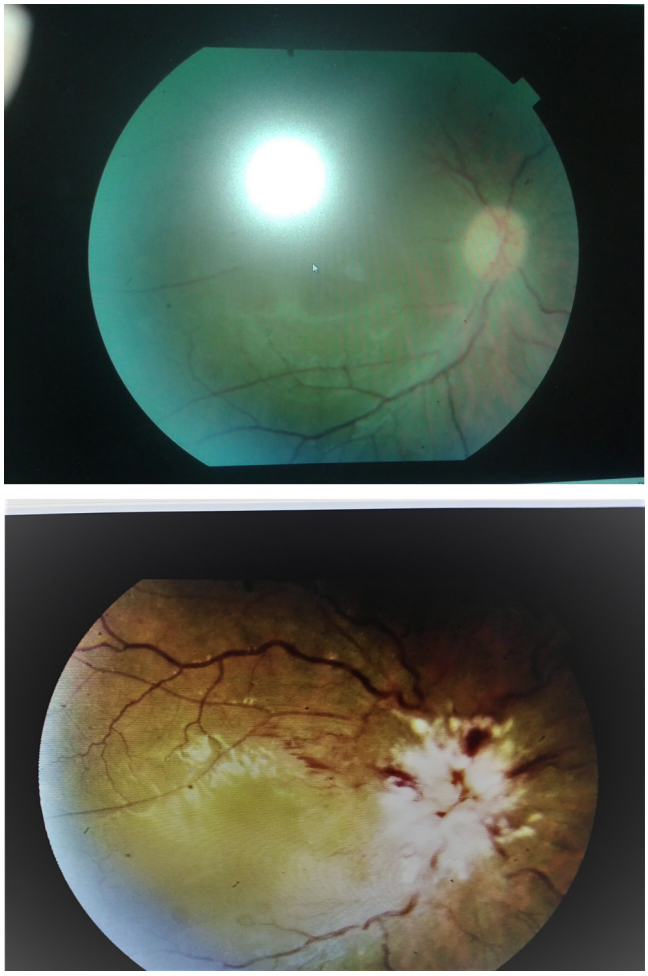

Fig. 2 – (a–b) Pre- and postoperative fundus photographs.

at the same rate as the general population, thus the association of pregnancy with PTC is probably only a measure of the fact that IIH affects women of child-bearing age [1]. We presented an unusual case of adult renal transplant recipient with IIH that was managed successfully with VP shunt. In this particular group of patients, early diagnosis and multidisciplinary care can salvage the patient from the risk of blindness associated with IIH.

Although no consensus exists on the best management strategy for IIH, effectiveness of acetolazamide, topiramate and octreotide have been assessed in several studies. Octreotide has been used in an uncontrolled open-label prospective study and the authors have reported that octreotide might be a promising therapeutic option for IIH. However, they have also expressed their questions which have remained to be answered in further clinical trials including the optimal treatment duration, the efficacy and safety of this drug in IIH [5].

It is well known that there is a direct link between obesity and IIH and that weight loss plays an important role in the management. In their study Altıokka-Uzun et al. have explained this link with a different aspect. Obesity is regarded as a proinflammatory state and is associated with increased expression of a number of adipokines and cytokines. They have noted that oligoclonal bands are also obtained in overweight individuals which is called ''rapid-onset obesity with hypothalamic dysfunction, hypoventilation and autonomic dysregulation syndrome'' [4].

However, despite loss of weight and best medical treatment strategies, surgical interventions become inevitable for patients with progressive visual loss or intractable headache. Procedures that are available include the placement of a lumboperitoneal (LP) shunt, VP shunt, optic nerve sheath fenestration (ONSF) or the insertion of a dural venous sinus stent [6,7]. The choice of decision depends on local expertise and the biases of the treating physicians.

The case presented here had an absolute indication for surgery due to the severe loss of vision and intractable headache. ONSF is a surgical technique in which optic nerve is exposed to several slits in dura and arachnoid mater [8]. However ONSF was not an appropriate choice for our patient as although might be effective in reversing vision loss, it does not provide any benefit for headache [9]. As intractable headache was amongst the major problems this surgical procedure was not considered as an option in our case. Other techniques which are evaluated under the heading of cerebrospinal fluid diversion techniques include lumboperitoneal (LP) shunt, ventriculoatrial (VA) shunt and VP shunt. Although investigators conclude that data is inconclusive to suggest or refuse any treatment modalities, it is reported that LP shunting is associated with higher complications when compared to VP shunting in addition to shorter survival rates [9]. On the other hand VA shunt is a technique which has been popular in the early 1950s [10]. However after experiencing serious complications and intraoperative difficulties VA shunts were generally avoided and consequently in the late 1970s, VA shunts became outdated following the introduction of VP shunt [11].

The literature regarding CSF diversion techniques are poor and inconclusive. There is only one study in the literature in which LP and VP shunt have been compared on the same symptoms management. In this study Tarnaris et al. have investigated the outcomes in patients with intracranial hypertension based on cerebrospinal fluid diversion site [12]. According to their results based on 34 patients, they have concluded that predicting which patients will improve is not possible and that the influence of site diversion is not critical but patients with VP shunt have less complications and revisions than those receiving a LP shunt.

Sinus stenting is an another efficient treatment option with low complication rates for cases with radiologically confirmed sinus venous thrombosis [13]. The only contraindication for transverse sinus stenting is dural venous thrombosis [13,14]. However no evidence for sinus venous thrombosis was evident in our case (Fig. 1b). Therefore VP shunt was the choice of treatment in our patient. Although placement of VP shunt in this patient population can often be difficult due to the small size of the ventricular system, intraoperative adjuvant techniques can be used to improve the accuracy and safety of VP shunt particularly for these patients. Various intraoperative adjuvant techniques have been employed in addressing the challenges of accurately cannulating the undersized ventricular system in IIH. These include the use of ultrasound guidance, neuronavigation, neuroendoscopy and intraoperative computed tomography (CT) [15]. In our case we used the

Table 1 – Surgical treatment options in PTC.

	Major indication(s)	Headache outcome	Visual outcome	Complications
ONSF[a]	Visual disturbances	Not indicated for headache	31–72%	Infection, transient or permanent diplopia, transient or permanent loss of vision, ischaemic optic neuropathy
LP shunt	Headache and visual disturbances	71%	42% resolution in papilloedema (15.9–22.7%)	Excessively low CSF pressure; obstruction of the catheter, infection
VP shunt[b]	Headache and visual disturbances	60%	40% resolution in papilloedema (100%)	Excessively low CSF pressure; obstruction of the catheter, infection
Sinus venous stenting[c]	Headache and visual disturbances	88%	87–91% resolution in papilloedema (91–97%)	Efficient technique with very low complication rates

[a] Optic nerve sheath fenestration.
[b] Ventriculoperitoneal shunt; revision rates are lower when compared to lumboperitoneal shunt.
[c] Indicated in cases those with radiologically confirmed sinus venous thrombosis only; contraindicated in dural venous thrombosis.

free hand technique for the placement of the ventricular catheter and confirmed the appropriate location on postoperative CT.

It is recommended that pregnant patients with IIH should be managed in the same way as any other patient with IIH. Treatment should based on management of preservation of vision and resolution of headache. Medical therapy includes carbonic anhydrase inhibitors (acetolazamide/topiramate), weight control, nonketotic diet, serial lumbar punctures, diuretics and analgesics [16]. Acetolazamide has double action, it works as a diuretic and it has been shown that CSF production reduces by 50% after acetalazomide treatment. However when medical treatment fails and/or not appropriate to use due to the reported of teratogenic risks in pregnant women, surgical interventions gain importance. Indications, contraindications in addition to outcomes regarding headache, vision loss and the resolution of papilloedema of the present surgery options for IIH was outlined in Table 1 [9].

Pregnancy is not considered to be a risk factor for IIH, however in pregnant patients with co-existing conditions such as being a recipient of renal transplant, IHH could be more prevalent [17–21]. Therefore, there should be a high index of suspicion for IHH in patients with persistent headaches and rapid visual loss. In refractory cases particularly in patients with co-existing conditions CSF diversion techniques could be the only exit to salvage vision and resolve the symptoms.

Conflict of interest

None declared.

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