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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.

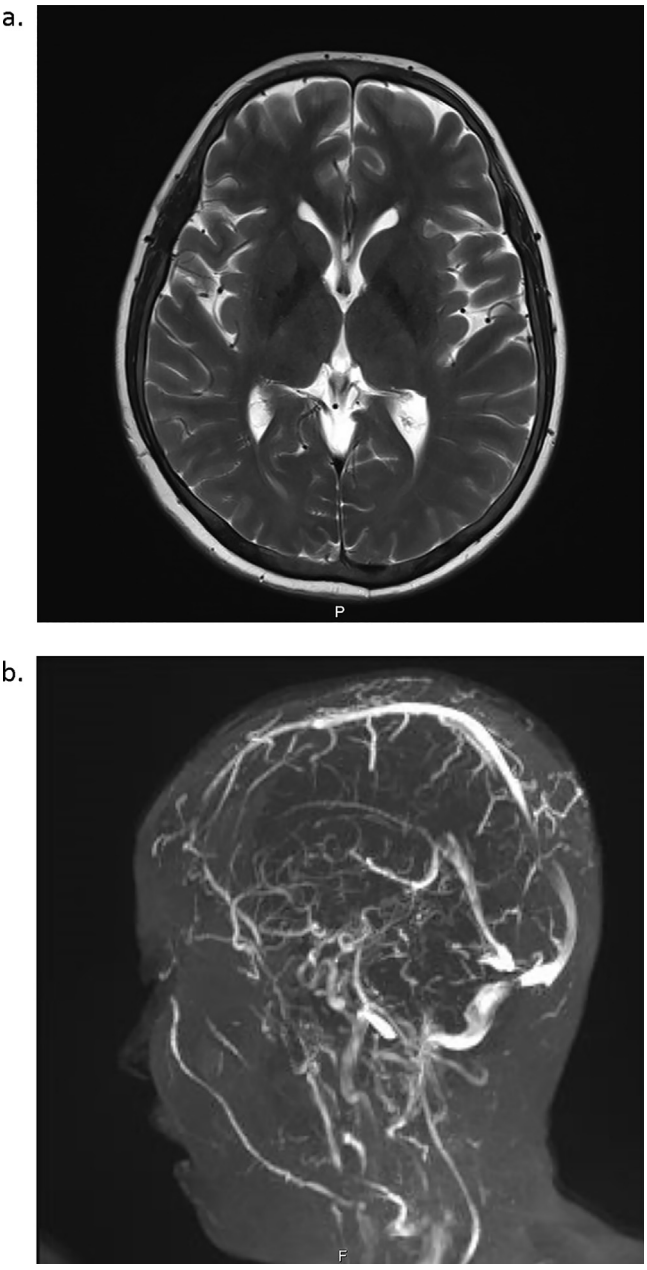

Fig. 1 – (a) Cranial MRI. (b) MR venography.

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

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