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Pseudotumour Cerebri in Pregnancy: An Antenatal Presentation

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Abstract

Pseudotumour Cerebri also called as benign intracranial hypertension is characterized by increased intracranial pressure, without a mass lesion and with elevated cerebrospinal fluid (CSF) pressure. Though its incidence in pregnancy is quite rare, the intractable headache associated and danger of impending visual loss warrants an early recognition, diagnosis and treatment.

The following is an antenatal presentation of benign or idiopathic intracranial hypertension or pseudotumour cerebri.

Keywords: *Pseudotumour Cerebri; Benign Intracranial Hypertension; IIH.*

Introduction

Idiopathic intracranial hypertension (IIH) also known as Pseudotumor Cerebri or benign intracranial hypertension is a syndrome of elevated intracranial pressure with normal CSF composition and no evidence of hydrocephalus or mass lesion. It is typically seen in obese women of childbearing age with an incidence of 19.3 per 100,000 [1].

During pregnancy the symptoms of IIH are similar to non-pregnant women and pregnancy in turn does not increase the risk of IIH [2]. Most common symptoms include headache, transient visual disturbances and pulsatile tinnitus. Fundosopic evaluation reveals progressive papilledema and sixth nerve involvement causing lateral rectus palsy and diplopia is also common. The role of serial ophthalmic evaluation is emphasized in literature to look for the presence of progressive worsening or improvement of disc edema and progression of field loss if any.

Pseudotumour cerebri is a diagnosis of exclusion, and is confirmed after all other causes on raised intracranial pressure like ICSOL or obstructive hydrocephalus are ruled out.

Case Report

A 23 year old patient at 24 weeks pregnancy, presented with intense headache, blurring of vision, diminution of vision and facial puffiness. Since the last two weeks of her presentation, her symptoms had progressively increased. She had no past history of similar complaints.

On examination her weight was 65 kg and her BMI was 24 kg/m^2. Her Blood pressure was within normal limits. She had facial puffiness but had no pedal edema.

An ophthalmological examination revealed normal visual acuity and normal anterior segment evaluation. Bilateral lateral react us palsy was noted with end gaze nystagmus. Fundus examination revealed bilateral disc edema. Fields 30-2 was within normal limits. Optic nerve sheath diameter was done using ultrasound B scan (right eye 4.2 mm; left eye 5.7 mm).

Her uterine height corresponded to her dates (24 weeks) and the rest of her obstetric examination was normal.

Her fundoscopy revealed papilloedema. Further investigation no proteinuria and essential normal blood investigations (Hb 10.4g/dl). An ultrasonograpy showed a single live intrauterine fetus of 23 to 24 weeks period of gestation. An essential diagnosis of pre eclampsia was ruled out.

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On MRI the following impression was made:

- Bilateral transverse sigmoid sinus stenosis,
- Buckling of bilateral optic nerve with distension,
- No evidence of any Intracranial space occupying lesion,
- Possibility of IIH (Idiopathic intracranial hypertension) to be considered.

Due to the intractable headache and signs of raised intracranial pressure, a CSF analysis and manometry was planned. The CSF analysis revealed the following report:

Protein: 23mg/dl, Glucose-65 mg/dl, Chloride-112 meq, Amylase-1.1, LDH-31.

The CSF pressure on manometry was**>** 535mmH20, this was suggestive of severe Idiopathic intracranial hypertension or pseudotumour cerebri. A diagnosis of benign idiopathic intracranial hypertension in pregnancy was confirmed.

A plan of management in collaboration with the neurologist, ophthalmologist and obstetrician was made. A therapeutic lumbar puncture was done and 80 ml of CSF drained.

In view of papilloedema and impending visual damage, tablet *Diamox* **(***Acetozolamide SR***)** 250 mg three times a day was started. A plan was made to increase acetazolamide to 1 gram per day gradually up to a maximum of 2g per day.

Within 48 hours of acetazolamide therapy it was seen that the patients headache resolved. On day 4, the torsional nystagmus resolved and on serial fundoscopy, it was noted that the papilloedema had markedly reduced as documented by ultrasound B scan.

Fig. 1: MRI showing absence of hemorrhage, infarct or cortical vein thrombosis

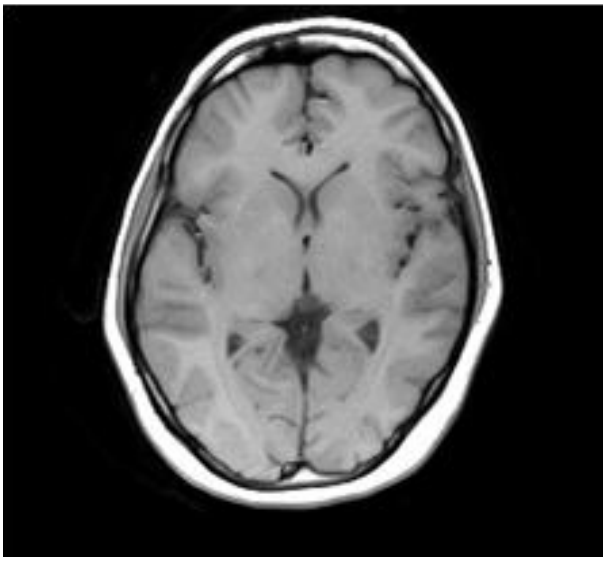


Fig. 2: Patent superior sinus and transverse sinus

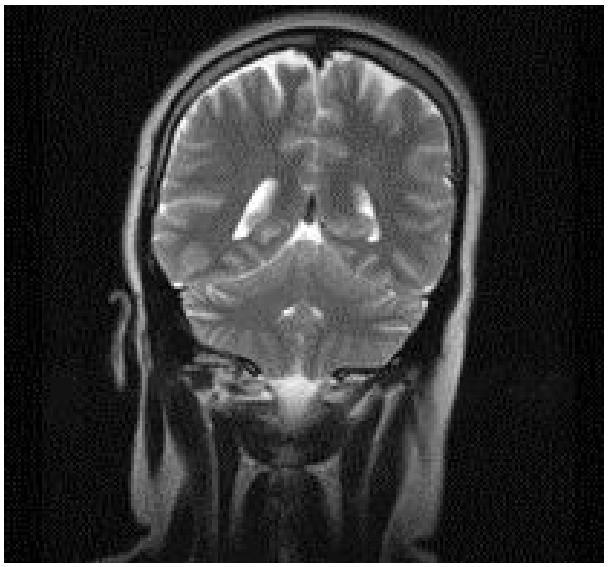


Fig. 3: MRI Venogram

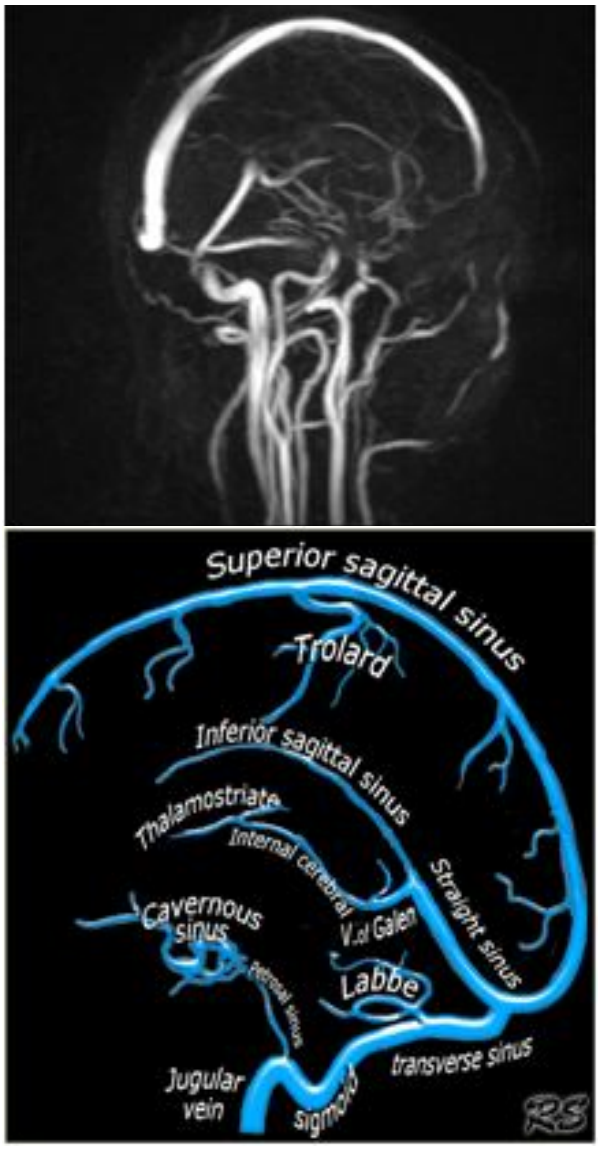


Fig. 4: Transverse sinus stenosis seen

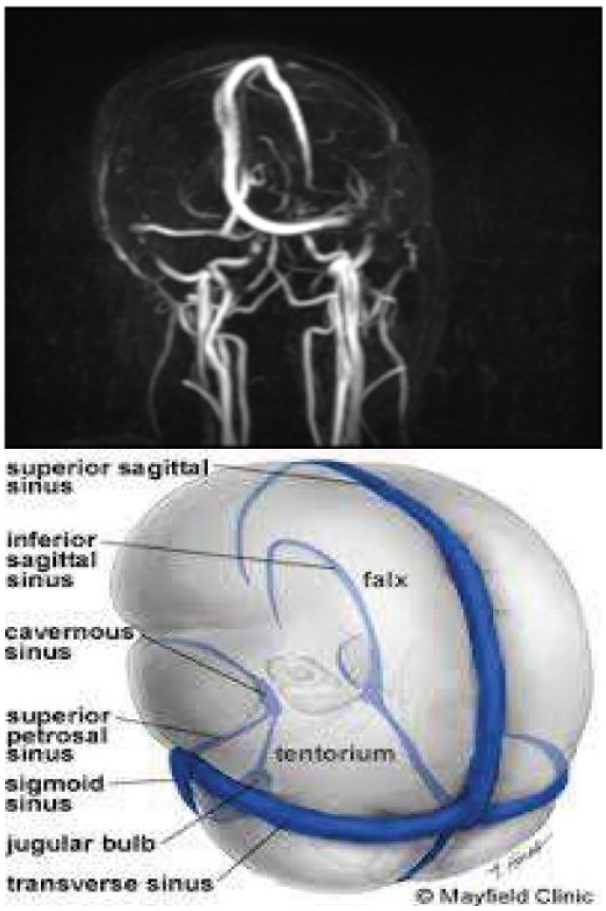
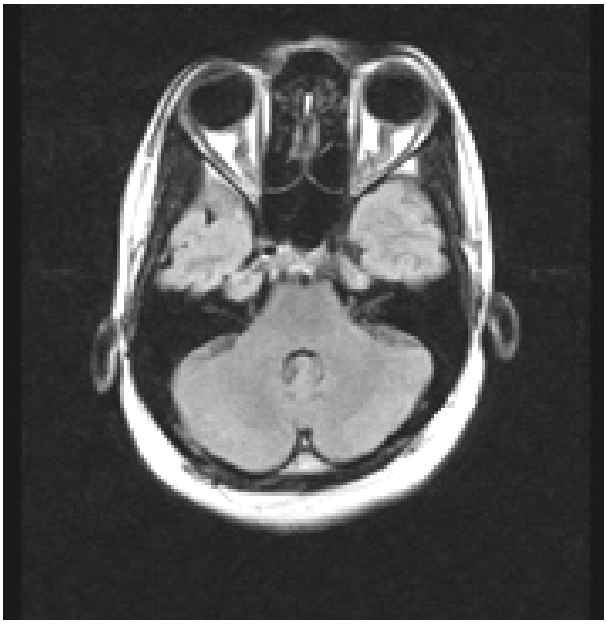


Fig. 5: Bilateral buckling of optic nerves seen



After the patient was asymptomatic, she was discharged one week later and maintained on acetozolamide 1g per day. She was advised follow-up visits every four weeks and at each visit the following was assessed: 1. Symptoms, 2. Visual field charting, 3. Optic nerve sheath diameter.

The patient progressed to 39 weeks of pregnancy with no headache or visual disturbances and resolving papilloedema, on acetazolamide 1g per day. Labor was induced at 39 weeks of gestation. She had an uneventful intrapartum and postpartum period and delivered a healthy full term baby of average weight.

Postnatally she was maintained on acetazolamide, which was gradually tapered and stopped.

Discussion

Pseudotumour cerebri also called as benign intracranial hypertension is defined by modified Dandy' s criteria [3]: increased intracranial pressure, without hydrocephalus or a mass lesion, with elevated cerebrospinal fluid (CSF) pressure but an otherwise normal CSF composition. The Incidence of IIH is: 19-24 per 1,00,000 obese women. Though essentially benign, 25% of patients develop permanent visual loss. The exact etiology for intracranial hypertension is not known, abnormal CSF absorption has been postulated as a cause.

In previous studies of pseudotumour cerebri in pregnancy, it has been seen that the rate of pregnancy and spontaneous abortions is not affected by this condition. It can occur in any trimester, and subsequent pregnancies do not pose a risk of recurrence.

The only difference in IIH in pregnancy exists with regard to the use of imaging and drugs. The goals of therapy are to preserve vision and to relieve symptoms.

The mode of delivery has to be planned depending on whether the patient has a previous history of shunt insertion. In all cases it is important to cut short the second stage of labor and avoid conditions that could further raise the intracranial tension. The plan of management to be followed ideally is:

- Headache therapy – Analgesics
- Serial Lumbar punctures
- Drug therapy: Acetozolamide (Category C)
- Surgical: Optic nerve sheath fenestrations and Lumboperitoneal shunts

Other medical treatment options that are indicated for IIH include furosemide, topiramate, and corticosteroids. Furosemide reduces intracranial

pressure by both diuresis and reducing the transport of sodium ions in the brain [4]. Like acetazolamide, it has been classified by FDA as the category C drug. The use of furosemide for IIH during pregnancy is recommended only when potential benefits justify the potential risks and that too for a short period of time [5].

Avoiding a rapid gain of weight in pregnancy, in obese antenatals, is an important part of management of IIH. Obesity causes a rise in central venous pressure and can exacerbate IIH.

Lumbar puncture (LP) is an effective management tool in pregnancy with IIH as it directly and immediately reduces intracranial pressure. Repeated spinal fluid drainage by LP can be helpful for improvement of symptoms of IIH in pregnancy and preventing permanent vision loss [2, 4].

LP is considered by some to be the 'treatment of choice' during pregnancy, and most patients experience improvement of symptoms for several days after a lumbar puncture, as in our case. Success of treatment was also seen on ophthalmoscopic exam, as the papilledema improved drastically.

In our case of intracranial hypertension in a pregnant patient, a fulminant course of progression was seen as compared to the usual protracted course. This case occurred de novo in pregnancy, with no prior history and patient had no pre-existing obesity.

Papilloedema was present at the time of presentation which made it a high risk case for impending visual loss. There was an involvement of the optic nerve and the abducens nerve involvement, which had resulted in diplopia.

When the mode of delivery and choice of anesthesia for an antenatal patient with IIH depends on whether she has had a CSF shunting procedure. Our patient was treated with lumbar puncture and acetazolamide and had a normal vaginal delivery. If a caesarean is warranted for obstetric indications, general anesthesia with its associated risks of aspiration and airway problems, especially in an obese parturient is best avoided. However, general anesthesia may be required in a patient with a lumboperitoneal shunt in place [6].

As an early diagnosis and suspicion of pseudotumour cerebri was present in our case, CSF drainage was able to provide immediate symptomatic relief and also preserve vision.

Conclusion

Pseudotumour cerebri although a rare entity should always be considered in the differential diagnosis of intractable headache in pregnancy associated with visual disturbances. A strong clinical suspicion, timely diagnosis accompanied by a prompt plan of management can go a long way in preventing permanent visual loss.

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