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Expert Opinion

Idiopathic Intracranial Hypertension in Pregnancy

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Quincke (who performed the first percutaneous lumbar puncture in 1890) first reported this condition in 1897 as "serous meningitis," which was later named, "pseudotumor cerebi" by Nonne in 1904. Idiopathic intracranial hypertension (IIH) has a female to male ratio of 8:1 and occurs in women of child-bearing age at a rate of about 1 in 100,000 in the general population but in almost 20 in 100,000 obese women with a mean age at the time of diagnosis of 30 years.

CASE

This is a 25-year-old G2 P1 female 20 weeks pregnant with a history of mild headaches about once a year. For the last 2.5 months, she has had a back of the head or bitemporal pressure and throbbing with an intensity of 9-10/10 with nausea, vomiting, light, and noise sensitivity, which has been fairly constant. She has taken no medication other than acetaminophen, which did not help. She has no infectious or systemic symptoms. She has gained about 30 pounds with the pregnancy.

She reports visual problems for about 1 month described as blind spots in the sides of vision of both eyes. The vision sometimes is blurry or blacks out for 2-3 seconds with standing. She has occasional ringing in the left ear but no hearing complaints.

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She saw an ophthalmologist and was found to have mild papilledema and enlarged blind spots on visual field examination. A magnetic resonance imaging (MRI) and venography (MRV) of the brain were normal. A lumbar puncture produced an opening pressure of 32 cm with a normal cerebrospinal evaluation except for a decreased protein level. Past medical history is negative. Her only medication is a prenatal vitamin. On examination, she is normotensive with a height of 5 feet 7 inches, and weight of 220 pounds. Neurological examination is normal except for the mild papilledema.

Questions.—How common is IIH (pseudotumor cerebri) during pregnancy? Is IIH a contraindication to a planned pregnancy? What treatments can be used during pregnancy? What about postpartum if the mother desires to breastfeed? Should labor and delivery or anesthesia be modified in any way? What is the prognosis of the pregnancy?

EXPERT COMMENTARY

Pseudotumor cerebri (PTC), also known as IIH, is a disease of obese young women of childbearing age. Pregnancy is a relatively common situation for these patients with PTC and Digre et al reported 28 pregnancies (21 patients) among 109 women with IIH. I believe that it is the weight gain associated with pregnancy that is the major risk factor for IIH in pregnancy but other hormonal or fluid related etiologies cannot be excluded.

In general, my evaluation and treatment of pregnant patients with IIH is the same as non-pregnant

patients with the exceptions: (1) I try to avoid contrast (Food and Drug Administration category C) in the MRI if I can in the first or second trimester; (2) I also would like to not use any medicines unless absolutely necessary especially for Category C agents like acetazolamide (Diamox) in the first trimester. If the patient needs medical treatment after the first trimester I consult with their obstetrician-gynecologist (OB-GYN) specialist and they generally allow the use of Diamox; (3) I offer serial lumbar punctures as a potential temporizing therapy through the first trimester for those who refuse medical or surgical therapy but have severe signs or symptoms; and (4) I recommend limiting the weight gain to the healthy normal and expected gain of pregnancy in coordination with their OB-GYN specialist.[1-5]

As with non-pregnant patients we check the systemic blood pressure (this patient is normotensive) and I ask about exogenous agents that can produce IIH (eg, excessive vitamin A including the prenatal supplements, steroids, tetracyclines, etc). I do tend to monitor these patients more closely than my normal IIH patients by seeing them in each trimester of their pregnancy if they are stable and I perform the typical IIH eye exam, fundus photography, and visual field testing. I also coordinate their headache management with their OB-GYN specialist (to select pregnancy appropriate therapies) and their neurologist. Some medications such as tricyclics can be considered after the first trimester but other headache medicines that are used in non-pregnant patients with IIH may be contraindicated in pregnancy.

My patients who have worsening but pre-existing PTC or who develop PTC during pregnancy undergo the same evaluation as my non-pregnant PTC patients, namely a MRI with MRV. Pregnant patients may develop cerebral venous sinus thrombosis from the hypercoaguable state of pregnancy and an MRI with MRV is my preferred initial imaging study for both pregnant and non-pregnant patients with IIH.

Contrast material is preferred in the evaluation of PTC and for the MRV but usually my radiologists will not give gadolinium in the first trimester as it is a category C agent. I explain the small risk of the procedure to the patient and some of my patients with pre-existing PTC choose not to undergo a repeat MRI or lumbar puncture (LP) at all and instead elect for observation or empiric treatment only. I tell these patients that after delivery if their symptoms and signs are not resolved then they will have to undergo the contrast MRI and MRV if they have chosen to undergo no imaging or LP at diagnosis.

As with non-pregnant patients, if patients decline, fail, are intolerant to, or are non-compliant with treatment and if there is progressive vision loss then surgery may be necessary. I prefer optic nerve sheath fenestration over shunting procedures because of the lower morbidity and mortality of sheath fenestration compared with shunt. In addition the peritoneal end of the shunt catheter may be affected by the growing uterus.

I do not consider IIH to be an indication for therapeutic abortion and I do tell the patients my bias in this regard. I also do not consider IIH in pregnancy to be a high risk pregnancy but close coordination of management with the OB is mandatory. I also defer to the patient and her OB-GYN the delivery mechanics but in general a spontaneous vaginal delivery is not felt to be of increased risk. Some authors believe that very high grade papilledema could potentially lead to visual loss from the Valsalva associated with labor and delivery but I generally do not recommend a Caesarean section or any change in the anesthesia regimen for my pregnant IIH patients. I do not tell young women considering a pregnancy who have preexisting IIH not to get pregnant but I do inform them of the potential risks of the weight gain of pregnancy and to contact us should they become pregnant. After delivery, I don't think that the patient should breast feed if they are on acetazolamide as it passes into the breast milk, but I defer the final decision to the patient and their OB-GYN. In my experience the prognosis of IIH in pregnant and non-pregnant patients is similar provided that they are monitored and counseled carefully.

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