

An Idiopathic Intracranial Hypertension Case with Rapid Improvement After Abortus and Review of the Literature

Abortus Sonrası Hızlı Düzelmeye Gösteren İdiyopatik İntrakraniyal Hipertansiyon Olgusu ve Literatürün Gözden Geçirilmesi

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ABSTRACT Idiopathic intracranial hypertension, also known as pseudotumor cerebri, is characterized by elevated intracranial pressure without hydrocephalus or a mass lesion. Idiopathic intracranial hypertension is most commonly observed in obese women of reproductive age, and it can also occur during pregnancy. Although weight gain and hormonal changes are common during pregnancy, the relationship between pregnancy and idiopathic intracranial hypertension is uncertain. Herein we report a pregnant patient that presented with headache, tinnitus, and diplopia. Neurological examination showed bilateral papilledema with bilateral sixth nerve palsy. Cerebrospinal fluid pressure elevated. The patient was diagnosed as idiopathic intracranial hypertension; her clinical symptoms resolved post abortion.

Key Words: Pseudotumor cerebri; pregnancy; papilledema

ÖZET İdiyopatik intrakraniyal hipertansiyon veya psödötümör serebri hidrosefali veya kitle lezyonu olmaksızın artmış intrakraniyal basınçla karakterize bir hastalıktır. İdiyopatik intrakraniyal hipertansiyon genellikle üreme çağındaki obez kadınlarda görülür. Ayrıca gebelikte de ortaya çıkabilir. Gebelikte kilo artışı ve hormonal değişiklikler sıklıkla görülmesine rağmen, gebelik ve idiyopatik intrakraniyal hipertansiyon arasındaki ilişki netleşmemiştir. Bu yazıda baş ağrısı, tinnitus ve diplopi yakınmaları ile başvuran, nörolojik muayenesinde bilateral papil ödem ve altıncı kraniyal sinir paralizisi saptanan hamile bir hasta sunuldu. Beyin omurilik sıvısı basıncı yüksek olarak bulunan hastaya idiyopatik intrakraniyal hipertansiyon tanısı konuldu. Hastanın klinik bulgularında abortus sonrası düzelmeye gözlemlendi.

Anahtar Kelimeler: Psödötümör serebri; gebelik; papil ödem

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Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri (PTC), is characterized by elevated intracranial pressure, without hydrocephalus or a mass lesion. IIH is most often seen in obese women of reproductive age.¹⁻¹⁰ IIH occurs in 0.8-1.7/100 000 in the general population, versus 19-24/100 000 among obese women of childbearing age.^{2,3,5,7} The etiology of the IIH is unknown, but numerous factors play a role, including endocrinological disorders, metabolic disorders, vitamin A deficiency, toxicity, and drugs (tetracycline, corticosteroids). Although weight gain and hormonal changes are common during pregnancy, the effect of pregnancy on IIH is uncertain. Prevalence studies have reported that there is no

increase in IIH during pregnancy; however, it was also reported that preexisting IIH could exacerbate during pregnancy and that IIH can occur in any trimester.^{2-8,11-13} Herein we report a case of IIH that was diagnosed during the fifth week of pregnancy in which the clinical symptoms resolved post abortion. To the best of our knowledge this is the first report of IIH with an onset as early as the fifth week of pregnancy that resolved so dramatically post abortion.

CASE REPORT

A 17-year-old female presented with a 2-week history of headache, tinnitus, and diplopia. Additionally, she reported gaining 15 kg during the previous 2 months. Physical examination was normal, but neurological examination showed bilateral papilledema and bilateral sixth cranial nerve palsy. There were no additional abnormal neurologic findings. The patient was overweight [Body mass index (BMI): 29.3] and her VAS pain score related to her headache was 8. Magnetic resonance imaging (MRI) and MR venography of the head were normal, and therefore disorders that can cause elevated intracranial pressure were excluded. Cerebrospinal fluid (CSF) pressure was high (330 mmH₂O) with normal cytological and biochemical findings. Her visual fields were severely constricted, with enlarged blind spots. Based on all the findings, she was diagnosed as IIH.

Routine hematological, biochemical and rheumatological tests revealed no abnormality. Hormonal blood tests showed an increase only in estrogen and β -hCG levels. β -hCG was also elevated based on subsequent measurements. Obstetric ultrasonography (USG) indicated a pregnancy of about 5 weeks. One week following the diagnosis of IIH she had an elective abortion after which her symptoms abated rapidly. One month post abortion her intracranial CSF pressure was 220 mmH₂O and she had lost 8 kg in body weight. Her fundus examination was normal, there was no papilledema, and visual field findings in the left eye were consistent with an enlarged blind spot. The patient was followed-up without treatment for 6 months during which time she was symptom free.

DISCUSSION

IIH (or PTC) was first described by Quincke.¹ The definition of IIH was then modified by Dandy based on the inclusion of radiological data. Since then, Modified Dandy criteria have been used to diagnose IIH.^{2,3} Although the clinical spectrum of IIH is very clear, the etiology of the disease is not precisely known. Most researchers agree that the disorder results from abnormal absorption of CSF.² Worrell et al. reported that increased extracerebral volume leads to an increase in intracranial pressure (ICP); however, this process progresses slowly, so that there is time for the brain's ventricular system to compensate, which explains why there is no dilatation of the cerebral ventricles in patients with IIH.²

Many factors, including metabolic, biochemical, hormonal abnormalities, and iatrogenic disorders, play a role in the development of IIH. In recent years researchers have focused on the etiological role of weight gain, as IIH is seen primarily in obese women, and have sought to develop treatment strategies accordingly. Central obesity causes intra-abdominal pressure to increase, which leads to an increase in pleural and cardiac filling pressures, subsequently causing a decrease in the venous return of the brain, and then finally an increase in intracranial venous pressure.³ Rapid weight gain is thought to be a very important factor in the development and treatment of IIH, and as such pregnancy is thought to be etiological or a factor associated with exacerbation of IIH.^{2-5,7,11-13} It was reported that 2%-12% of pregnant women have IIH.^{3,5} IIH can occur in any trimester of pregnancy, and in patients previously diagnosed as IIH pregnancy can exacerbate its symptoms.^{2-6,8} The clinical course of IIH does not differ between pregnant and non-pregnant patients. Thus, visual field defects and visual loss is the most important cause of morbidity; visual outcome is similar in both pregnant and non-pregnant patients.^{3-5,7-9}

The rate of spontaneous abortion in IIH patients is the same as in the general population, so therapeutic abortion is not indicated to limit pro-

gression of the disease.^{2-5,8} Additionally, fertility treatment in patients with IIH is not contraindicated; however, if prenatal vitamins are used they should be checked for vitamin A content.⁶ In patients with IIH the management of labor should be the same as in normal pregnancies; however, in cases of Cesarean delivery epidural anesthesia should be used instead of general anesthesia, which might be associated with an increase in CSF pressure.^{3,4,8} In general, subsequent pregnancies do not increase the risk of recurrence, although recurrent IIH in pregnancies has been reported.^{3-6,8}

The goal of the treatment and follow-up of IIH is to preserve vision and improve symptoms, as vision loss occurs in 10% of pregnant women with IIH.^{2,3} As weight loss has been associated with symptom improvement in IIH, the treatment plan for pregnant patients must include careful weight management while remaining aware of ketosis.^{2-6,8} Bed rest and acetaminophen for treatment of headaches are recommended.² Acetazolamide, a carbonic anhydrase inhibitor, is the first line treatment in IIH, but it may be teratogenic in pregnancy; however, there are some reports on the use

of acetazolamide in pregnant IIH patients.^{3,5} Among these studies, the number of patients that used acetazolamide during pregnancy and did not experience any problems were in a retrospective study reported by Huna-Baron et al., in a study by Digre et al., and in a study by Lee et al.^{5,9,11} Bagga et al. reported a case in which they performed abortion upon the patient's request and observed improvement in symptoms, as in the presented case.³ Steroids should be used only in cases of progressive vision loss. Visual field tests should be performed regularly and if there is unpreventable vision loss surgery should be considered as an option. Optic nerve fenestration and lumboperitoneal shunt placement can be performed during pregnancy if necessary.²⁻⁸

In conclusion, IIH can occur during the very early phase of pregnancy, as in the presented case. Rapid improvement post abortion and exacerbation of symptoms after weight gain are indicative of the role of pregnancy in the development of the disease. As such, careful management of weight should be considered during the treatment of pregnant IIH patients.

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