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NARRATIVE REVIEW

Multidisciplinary management of idiopathic intracranial hypertension in pregnancy: case series and narrative review

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Abstract Idiopathic intracranial hypertension (IIH) is a neurological condition characterized by raised intracranial pressure of unknown etiology with normal cerebrospinal fluid (CSF) composition and no brain lesions. It occurs in pregnant patients at approximately the same frequency as in general population, but obstetric and anesthetic management of the pregnancy and labor remains controversial. In this article we provide a multidisciplinary review of the main aspects of IIH in pregnancy including treatment options, mode of delivery and anesthetic techniques. Additionally, we report three cases of pregnant women diagnosed with IIH between 2012 and 2019 in our institution.

Introduction

Idiopathic intracranial hypertension (IIH) is a neurological condition with a benign course characterized by raised intracranial pressure of unknown etiology. In these patients, the cerebrospinal fluid (CSF) composition is normal and brain lesions are absent.[1] It is a rare condition, with an estimated incidence of 0,9 per 100,000 population.[1] It occurs in pregnant patients at approximately the same rate as in general population.[1] During pregnancy it generally appears in the first half of gestation although IIH can appear in any trimester of pregnancy and pregnancy does not appear to alter the natural history of the disease.[1,2] A multidisciplinary evaluation of this patients during pregnancy and labor is essential. We will review the main aspects of IIH, including the obstetric and anesthetic considerations in the parturient with IIH, and report three cases that occurred in our institution between 2012 and 2019.

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Table 1 Modified Dandy criteria for idiopathic intracranial hypertension.

Modified Dandy criteria for idiopathic intracranial hypertension
1. Signs and symptoms of increased intracranial pressure
2. Absence of localizing findings on neurologic examination
3. Absence of deformity, displacement, or obstruction of the ventricular system and otherwise normal neurodiagnostic studies, except for evidence of increased cerebrospinal fluid pressure
4. Awake and alert
No other cause of increased intracranial pressure present

Pathogenesis

The pathogenesis of IIH remains unclear but proposed etiologies suggest that it is caused by accumulation of CSF due to a defect in arachnoid villi reabsorption. An increased CSF production, cerebral edema, and abnormalities in cerebral blood flow (e.g. venous stenosis or venous hypertension) seem to be also involved.[2,3] Obesity may play a role through changes in sodium and water retention mechanisms, and also by increasing abdominal pressure which increases pleural and cardiac filling pressures, delaying venous return from brain resulting in increased intracranial venous pressure.[3–5]

Pregnancy was previously reported as an etiologic factor for IIH and the hyperestrogenemia, along with thrombophilia and hyperfibrinolysis, characteristic of pregnancy, were proposed as mechanisms that could promote or worsen IIH.[4] Nevertheless, this association was not clearly established.

Clinical presentation

The most frequent symptom of IIH is a generalized headache exacerbated with Valsalva maneuver and eye movement, being more severe in the morning. However, the features of headaches are variable and are not specific to IIH. It may be accompanied by photophobia, neck and back pain, nausea, vomiting, and tinnitus. Visual disturbances are common and IIH may present with diplopia, loss of acuity, or visual field.[4,5] The physical exam reveals papilledema, that is the hallmark sign of IIH, and it is usually bilateral and symmetric. Visual loss is the major morbidity in IIH and commonly gradual, but when its onset is abrupt and if intracranial hypertension is untreated it may cause permanent visual loss.[5]

Diagnosis

Idiopathic intracranial hypertension is a diagnosis of exclusion, so secondary causes must be excluded. The diagnosis is based according to Modified Dandy criteria for IIH (Table 1).[1,6]

Neuroimaging is required to exclude secondary causes of intracranial hypertension.[1,3] Magnetic Resonance Imaging (MRI) is safe and is the method of choice during pregnancy. When no structural or vascular lesion is identified it should be followed by lumbar puncture (LP).[5,7]

LP is an essential element to establish the diagnosis of IIH, defined as an opening CSF pressure above 25 mmHg. The evaluation of CSF contents must be normal to define IIH.[5,8] Ophthalmologic evaluation is imperative to evaluate the severity of optic nerve involvement and monitor response to treatment.[9]

Treatment

There are two major goals in treating IIH which are improvement of symptoms, predominantly headaches, and the preservation of vision. In general, pregnant women can be treated as nonpregnant adults, although with some considerations.[4,5]

Weight control is very important and a low-calorie diet should be started.[2] Considering that this approach can take some time to achieve effective outcomes and that excessive weight loss can induce adverse effects on the fetus (e.g. ketosis), other treatments should be tried simultaneously.[4,10]

Acetazolamide, a carbonic anhydrase inhibitor, reduces cerebrospinal fluid production and is the first line medical option for IIH in adults. However, its use in pregnant women remains controversial due to several reports of teratogenic effects in animals and a single case of a sacrococcygeal teratoma in humans. Food and Drug Administration classifies acetazolamide as a class C in pregnancy, even though there is a lack of adequate controlled studies in pregnant women and little clinical evidence that supports any adverse effects of this drug.[3] The use of other diuretics is usually not recommended during pregnancy because the lowering of maternal blood volume can reduce placental blood flow.[5,11,12] Corticosteroids should be reserved for acute visual loss situations.[3,5,13] Serial lumbar punctures can transiently relieve symptoms since CSF reforms within six hours. Furthermore, lumbar punctures can be painful, technically difficult in obese and pregnant women, and complicate with CSF leak or infection. Nonetheless, this is the preferable approach in many institutions during pregnancy.[2,5,13]

Surgical treatment is reserved for patients with severe progressive visual loss or persistent headache despite optimal medical therapy.[1] Optic nerve sheath fenestration option seems to be more beneficial to visual function, and lumboperitoneal or ventriculoperitoneal shunt can be technically difficult in pregnant women due to the gravid uterus.[5,14]

Management of pregnancy and labor

There is no indication to terminate a pregnancy in a woman diagnosed with IIH because gestation does not worsen the prognosis of IIH, neither affects perinatal outcome.[3,5]

Mode of delivery is often a controversial decision when a pregnant woman presents with IIH.[5,8]

Physiologic changes in pregnancy could change intracranial pressure. The increase in blood volume and cardiac output, combined with increased water and sodium retention, promote a progressive increment in cerebral blood flow, possibly causing cerebral edema. Despite this changes, CSF pressure is unaltered (7–15 mmHg) in normal pregnancy.

However, during the first and second stages of labor CSF pressure can rise to 39 and 71 mmHg, respectively.[1]

The concern is based on the theory that pushing efforts and uterine contractions increase blood pressure, cardiac output, and central venous pressure, consequently increasing CSF pressure. Nonetheless, an instrumented delivery – vacuum, forceps, or spatulas – is a good option to reduce maternal pushing efforts on the second stage of labor and thereby reducing the potential increase in CSF pressure.[5,8] IIH is not considered an indication for an elective cesarean delivery.[5,15]

Anesthetic considerations

Labor analgesia and cesarean anesthesia are a challenge to the anesthesiologist. The main goal is maintaining hemodynamic stability in order to control cerebral perfusion pressure and brain tissue oxygenation. Increases in intracranial pressures and abrupt decreases in mean arterial pressures must be avoided.

The anesthetic choice for IIH patients is complex and depends on balancing the risks and benefits of each available technique.[16,17] Although neuraxial anesthesia is contraindicated in patients with intracranial hypertension resulting from space occupying lesions due the risk of uncal herniation, in IIH patients there is a uniform swelling of the brain that prevents herniation, so neuraxial anesthesia can be used safely.[1,16,18,19] Spinal anesthesia will increase the volume of fluid in the subarachnoid space and epidural anesthesia will compress the dural sac, altering the compliance of spinal subarachnoid space.[20,21] There are case reports of successful use of both spinal and epidural anesthesia for cesarean delivery in IIH patients.[22–24]

Spinal anesthesia alone or combined with epidural has been used safely in IIH patients. It is crucial to use small volumes of local anesthetic and opioids in order to avoid an acute rise in intracranial pressure.[16] It allows CSF drainage and the use of small volumes of local anesthetic. The placement of a spinal catheter permits the monitoring of ICP.[25] The hypotension associated with spinal anesthesia reduces cerebral blood flow and cerebral perfusion pressure, therefore fluid load and vasoactive drugs should be available in order to minimize this risk. The anesthetist should closely monitor hemodynamic stability and neurological signs. An epidural catheter can be used with precaution due to the increase in epidural volume that will be transmitted to the subarachnoid space, increasing the intracranial pressure transiently. The rate of injection should be as slow as possible. Slowly incremental doses seem to be better tolerated than a high-volume dose.[21] Neurological, cardiovascular and respiratory monitoring should be prolonged in the next hours after the procedure.

General anesthesia in pregnancy is associated with several risks, including difficult airway, aspiration, awareness, and potential masking of neurological changes in IIH patients. In these patients, general anesthesia should also be avoided since laryngoscopy, intubation, inadequate depth of anesthesia, and extubation are associated with a significant raise in intracranial pressure.[16,18,20] If general anesthesia is necessary, it should be planned carefully to avoid intracranial pressure variations. In these cases, pharmacological choices are essential. Propofol is an intravenous induction agent that offers the advantage of decreasing cerebral blood flow, protecting the brain tissue.[18,21] The use of opioids is controversial and they should be carefully selected and titrated to avoid potential neonatal respiratory depression. Concerning neuromuscular blocking drugs, succinylcholine should be avoided for intubation because muscle fasciculations may raise intracranial pressure transiently.[18] The depth of anesthesia should be monitored. Extubation should be performed in a deep plain of anesthesia.[20,21] Mechanical ventilation should be carefully controlled with tight adjustment of carbon dioxide arterial pressure, in order to minimize its effects on cerebral blood flow.

Cases reports

Case 1

A 21-year-old multiparous woman at 18 weeks pregnant presented with frontal headache, nausea, and dizziness with 3 days of evolution. She was overweight, had a history of migraine, and smoking habits. On physical examination she had bilateral asymmetric papilledema but visual fields, acuity, and head MRI were normal. Diagnostic LP showed an opening pressure of 29 mmHg and a normal composition of CSF. In this LP 9 mL of CSF were drained. The severity of headache improved but she noted additional visual symptoms, like blurred vision. A second LP puncture was necessary in order to improve symptoms. After a multidisciplinary discussion, including obstetricians, neurologist, and anesthesiologist it was decided to terminate pregnancy at 38 weeks with an elective cesarean section to prevent acute relapse of intracranial hypertension. On presentation to the cesarean she was asymptomatic. Monitoring included pulse oximetry, electrocardiogram, noninvasive blood pressure, and urine output. A spinal anesthesia was selected. A 26G Quincke needle was used and 9 mg hyperbaric bupivacaine (5 mg.mL^{-1}) and 0.015 mg fentanyl were slowly injected. Multimodal analgesia was provided with 1000 mg intravenous paracetamol, 200 mg intravenous tramadol and 75 mg intramuscular diclofenac. The procedure was uneventful.

After delivery she had persistence of headache and intracranial hypertension symptoms, with no effect on visual fields, that were treated with acetazolamide 500 mg twice daily and two more CSF drainage with LP. With this approach there was a successful improvement of symptoms.

Case 2

A 30-year-old nulliparous, smoker, and obese woman with 18 weeks of gestation, presented at the emergency service with transient visual obscurations and tunnel vision with 3 weeks of duration but no headache. On physical examination she had bilateral papilledema but visual fields and acuity were normal with preserved hemodynamic stability. MRI revealed an empty sella turca image with enlargement of optic nerve dural sheaths. The first LP showed a CSF opening pressure of 47 mmHg with normal biochemical and cytological composition. At this stage, intracranial hypertension was managed with corticosteroids (methylprednisolone 250 mg once daily). The patient reported rapid improvement of

symptoms. After this episode, pregnancy was managed with a dietary weight control plan and four serial CSF drainages with lumbar punctures, showing a progressive decreasing opening CSF pressures. With this approach the patient noted an improvement of visual symptoms, with no headache history.

A cesarean section was scheduled in order to prevent intracranial hypertension exacerbations. On presentation for cesarean she was asymptomatic. Monitoring included pulse oximetry, electrocardiogram, noninvasive blood pressure and urine output. A spinal anesthesia was chosen. A 27G Quincke needle was used to withdraw passively 3 mL of CSF and then anesthesia was initiated with intrathecal 8 mg of hyperbaric bupivacaine (5 mg.mL^{-1}) and 0.002 mg sufentanil. The cesarean occurred uneventfully. Multimodal analgesia was provided with intramuscular diclofenac 75 mg. All symptoms and papilledema resolved on postpartum period, with no more treatment needed. No perinatal adverse outcomes were documented.

Case 3

A 27-year-old multiparous woman with excessive weight presented at the emergency service with a six-month history of holocranial headache that worsened at night with a refractory response to analgesia and progressive visual symptoms (visual obscurations and loss of vision on left eye hemicamp). Physical examination revealed altered visual fields and optic nerve atrophy, as well as a discrete decrease on right eye visual acuity. MRI showed a prominence of the suprasellar cistern and enlargement of optic and oculomotor nerve dural sheaths, changes of idiopathic intracranial hypertension. LP revealed an opening CSF pressure of 37 mmHg, and 30 mL were drained. Cytological and biochemical CSF analysis were normal. Acetazolamide 500 mg twice daily was started, with marked improvement of symptoms. In the meantime, the patient discovered that she was pregnant with 25 weeks of gestation. After a multidisciplinary discussion, it was decided to stop acetazolamide due to the potential teratogenic risks. Two serial LP were performed, showing opening pressures of 23 mmHg in both occasions.

Patient remained asymptomatic for the rest of the pregnancy and elective cesarean was scheduled at 39 weeks. On presentation for cesarean she was asymptomatic. An epidural anesthesia was used. The epidural space was located at the L3-4 interspace with the patient in lateral decubitus using an 18G Tuohy needle with loss-of-resistance to air. Anesthesia was provided through the epidural catheter with 75 mg ropivacaine (7.5 mg.mL^{-1}), and 0.01 mg sufentanil. A satisfactory level of block was achieved and cesarean occurred uneventful. Multimodal analgesia was provided with intravenous paracetamol 1000 mg and intramuscular diclofenac 75 mg. At the end of the surgery, the epidural catheter was removed. There were no symptoms, neurologic changes or other complications.

No fetal malformations were detected and no complications reported in perinatal period.

On maternal postpartum evaluation she reported only occasional mild headaches but visual fields remained altered and fundoscopy showed persistent optic nerve atrophy with no papilledema.

Discussion

The management of pregnancy and delivery in pregnant women with IIH is complex and controversial. Serial lumbar punctures can be part of the management of these patients and were the treatment of choice for the three cases presented in this article.

Although many review articles on IIH suggest that acetazolamide should be avoided in pregnancy, there is paucity of clinical evidence for this recommendation.[8,11] There was only a single case of sacrococcygeal teratoma reported in an infant of a mother treated with acetazolamide during first half of the pregnancy, in 1978.[26,27] There are no well-documented reports of adverse fetal effects of acetazolamide used during pregnancy. Therefore, it is important to promote a multidisciplinary approach involving neurologist and individualize each case, providing a careful risk-benefit assessment regarding the use of acetazolamide.[28,29] It might be considered if the risk of progressive visual loss outweighs potential risks.

Management of labor and mode of delivery are also controversial. In our report, all three cases underwent cesarean delivery based on the assumption that uterine contraction and bearing-down efforts during vaginal delivery could increase CSF pressure. However, studies suggest that IIH is not itself a specific indication for cesarean delivery.[5,15] The rise on CSF pressure is transient and vaginal deliveries have been reported with no adverse effects. Additionally, there is no evidence that either mode of delivery is superior in these patients, so the recommendation is that de decision should be based on obstetric indications and not dependent on the presence of IIH.[5,15]

The decision regarding the choice of the anesthetic technique for labor or cesarean should be individualized and discussed with the team because there are no published randomized controlled trials comparing the safety of neuraxial *versus* general anesthesia. The main goal is to avoid increase in ICP, using regional techniques or general anesthesia.

In 1979, Palop et al. reported two cases of lumbar epidural for labor analgesia in IIH patients.[30] Later, Perez Rodriguez reported the use of an epidural catheter for cesarean anesthesia and postoperative analgesia.[19] Moore and colleagues and Guerci et al. also reported cases of effective use of epidural anesthesia in IIH patients.[14,16] A successful use of combined spinal-epidural techniques was reported for Bedson and Plaat in IIH patient for cesarean.[23]

Intrathecal catheters are also an option for the management of these patients. Aly reported the use of an intrathecal catheter in labor analgesia and Moore et al. used an intrathecal catheter for cesarean anesthesia.[16,24]

In 2016, Gragasin and Chiarella reported a case of IIH in which the first option was an epidural catheter, but an unintended dural puncture occurred and an intrathecal catheter was inserted and used for labor analgesia, removal of CSF, and cesarean anesthesia.[25]

General anesthesia has also been reported as a safe option and it was the choice of Aboulish and colleagues for cesarean in a patient diagnosed with IIH.[31]

We report safe approaches to neuraxial techniques. Spinal anesthesia with or without LCR drainage can be performed using small volumes of local anesthetics and was

the technique of choice in two cases of our institution. In the third case, we decide to perform an epidural anesthesia using slow rate of injection in order to minimize the transmission of pressure to subarachnoid pressure.

Although we prefer neuraxial approach for IIH cases in our institution, there are reported cases of safe use of general anesthesia for cesarean in these patients.[21,31] General anesthesia in parturient have several risks of difficult airway, aspiration, and awareness. In cases of IIH, general anesthesia makes impossible to detect alterations on mental status that can be indicative of increasing ICP.

In conclusion, it is essential an antenatal multidisciplinary consultation to discuss the obstetric management and the anesthetic choice in order to promote an optimal and individualized approach to each case of IIH.

Conflicts of interest

The authors declare no conflicts of interest.

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