A case report of pregnancy complicated by Idiopathic Intracranial Hypertension and bilateral papilledema

Dr. Rakshitha. S[1]*, Dr. Ramya. S[2]

[1,2] Department of Obstetrics and Gynaecology, Sri Ramachandra Institute of Higher Education and Research, Porur, Chennai, Tamil Nadu, India

ABSTRACT:

Raised intracranial tension (ICT) during pregnancy is rare but potentially life-threatening, requiring prompt diagnosis and multidisciplinary management. Papilledema, a hallmark of increased ICT, can pose significant maternal and fetal risks, especially in late gestation. We report a case of a 32-year-old primigravida at 37+2 weeks of gestation, conceived by intrauterine insemination (IUI), with hypothyroidism and gestational diabetes mellitus (GDM). She presented with frontal and periorbital headache. Ophthalmic evaluation confirmed bilateral early papilledema secondary to raised ICT. She had been on oral acetazolamide and sodium bicarbonate therapy since mid-pregnancy for the same. Multidisciplinary evaluation (neurology, nephrology, ophthalmology, anesthesiology) was undertaken, and an emergency lower segment cesarean section (LSCS) was performed under general anesthesia. A healthy female neonate was delivered. The postoperative course was uneventful, and the patient was discharged on day 7 with continuation of neurological medications. Thus, increased ICT with papilledema during pregnancy necessitates careful balancing of maternal neurological safety and fetal maturity and multidisciplinary coordination enables favorable outcomes.

Keywords : **Papilledema, Raised intracranial tension, Acetazolamide, Pregnancy**

INTRODUCTION:

Raised intracranial tension (ICT) in pregnancy is uncommon and may be caused by idiopathic intracranial hypertension (IIH), intracranial space-occupying lesions, cerebral venous sinus thrombosis, or other neurological disorders. The hallmark clinical sign is papilledema, which can be detected during ophthalmologic examination. Untreated, increased ICT may result in permanent visual impairment, herniation syndromes, or maternal-fetal morbidity. Pregnancy complicates management due to physiological changes, limited drug safety data, and the need to time delivery appropriately. Acetazolamide, a carbonic anhydrase inhibitor, is commonly used for IIH but requires careful risk-benefit analysis in pregnancy. We present a case of bilateral early papilledema secondary to raised ICT in late pregnancy, managed successfully with emergency cesarean section after multidisciplinary planning.

*Corresponding Author

CASE REPORT:

A 37 weeks +2 days primigravida presented with a one-day history of insidious-onset frontal and periorbital headache. The headache had no clear aggravating or relieving factors. There was no history of vomiting, seizures, visual disturbances, limb weakness, or altered consciousness. The antenatal course had been largely uneventful. First-trimester nuchal translucency and screening tests were low risk, and the anomaly scan was normal. She received two doses of tetanus toxoid and was compliant with iron and calcium supplementation. In the second trimester, she was diagnosed with gestational diabetes mellitus based on oral glucose tolerance test and was managed with medical nutrition therapy. At 22 weeks gestation, an ophthalmological evaluation revealed bilateral early papilledema. Following a neurology consultation, she was initiated on acetazolamide and sodium bicarbonate, which were continued under nephrology guidance. Third trimester was uneventful, growth scan showed fetal growth appropriate for centiles and patient perceived fetal movements well.

Patient was admitted in view of insidious-onset frontal and periorbital headache. On examination at presentation, her vital signs were stable with a blood pressure of 120/80 mmHg, heart rate of 84/min and a respiratory rate of 17/min. General physical examination revealed no pallor, icterus, cyanosis, clubbing, lymphadenopathy, or pedal edema. Systemic examination showed no abnormalities in the cardiovascular and respiratory systems. Neurologically, she had no focal deficits. Obstetric examination revealed a term uterus with a cephalic presentation (5/5th palpable), clinically adequate liquor, and regular fetal heart sounds.

Laboratory investigations showed hemoglobin of 12.0 g/dL, total leukocyte count of 8,920/cumm, and platelet count of 1.79 lakh/cumm. Serum bicarbonate was 16 mmol/L, suggesting metabolic acidosis attributed to acetazolamide use. Repeat ophthalmological assessment confirmed persistent bilateral early papilledema, and both neurology and nephrology teams advised continuation of the current medical regimen. Given the presence of raised intracranial tension (ICT) with persistent headache at term gestation, a multidisciplinary team decision was made to proceed with an emergency lower segment cesarean section (LSCS). The LSCS was under general anesthesia via a Pfannenstiel incision. Intraoperatively, the lower uterine segment was not formed. A live female baby weighing 2.73 kg was delivered with clear liquor, with APGAR scores of 8 and 9 at one and five minutes respectively, and delayed cord clamping was performed. The placenta was delivered intact without any extension or postpartum hemorrhage. The uterus was closed in two layers and hemostasis was secured and estimated blood loss was 450 ml.

Postoperatively, the patient was hemodynamically stable and neurology advised restarting acetazolamide at 125 mg twice daily. On POD 2, nephrology advised restarting sodium bicarbonate at 1 gm three times daily. On POD 3, the surgical wound was healthy and the dressing was removed. On POD 7, sutures were removed, and both mother and baby were discharged in stable condition with appropriate follow-up instructions.

DISCUSSION:

Pathophysiology of Idiopathic intracranial hypertension in pregnancy :

Idiopathic intracranial hypertension (IIH) in pregnancy represents a complex interaction between the baseline pathophysiology of IIH and the unique physiological changes of gestation. IIH is defined by elevated intracranial pressure (ICP) in the absence of a mass lesion, hydrocephalus, infection, or abnormal cerebrospinal fluid (CSF) composition. The core pathological process is believed to be impaired CSF absorption at the level of the arachnoid villi and granulations, leading to accumulation of CSF and raised ICP. This may be due to structural abnormalities, inflammation, or secondary effects of increased cerebral venous pressure. MR venography in many patients demonstrates transverse venous sinus stenosis, which may be either a primary anatomical narrowing causing venous hypertension or a secondary, reversible compression caused by the elevated ICP itself, creating a self-perpetuating cycle.

During pregnancy, normal physiological changes can exacerbate these mechanisms. Maternal plasma volume expansion, increased cardiac output, and hormonal effects on sodium and water retention lead to higher intracranial blood volume. Elevated progesterone and estrogen alter vascular tone and permeability, potentially affecting cerebral venous drainage. The pregnancy associated hypercoagulable state increases susceptibility to cerebral venous sinus thrombosis which is a critical differential diagnosis that must be excluded before diagnosing IIH. Furthermore, pregnancy-related weight gain can worsen the pre-existing obesity-linked risk factors for venous congestion and impaired CSF resorption. The net result in pregnancy is a heightened vulnerability to ICP elevation, with symptoms ranging from headache and transient visual obscurations to progressive visual field loss due to papilledema. Despite these risks, the underlying brain parenchyma remains structurally normal, and the condition is potentially reversible with timely diagnosis and intervention.

Idiopathic intracranial hypertension (IIH) is diagnosed based on the Modified Dandy criteria as refined by Friedman et al. (2013), all of which must be fulfilled. The condition presents with symptoms and signs of raised intracranial pressure such as diffuse daily headaches, transient visual obscurations, pulsatile tinnitus, diplopia (often due to sixth cranial nerve palsy), and papilledema, which is the most common clinical finding. Neurological examination typically reveals no localizing deficits, except for possible unilateral or bilateral abducens nerve palsy. Neuroimaging, preferably magnetic resonance imaging (MRI) with magnetic resonance venography (MRV), is performed to exclude secondary causes such as mass lesions, hydrocephalus, or venous sinus thrombosis, and may show supportive signs including flattening of the posterior sclera, distension of the perioptic subarachnoid space, empty sella, and transverse sinus stenosis. Cerebrospinal fluid (CSF) analysis demonstrates normal composition with no evidence of infection, inflammation, or malignancy. Lumbar puncture reveals an elevated opening pressure—defined as ≥ 250 mmH_2O in adults, or ≥ 280 mmH_2O in children, measured in the lateral decubitus position with legs extended and the patient relaxed. In pregnancy, the same criteria apply, though MRV without contrast is preferred to rule out cerebral venous sinus thrombosis. Lumbar puncture is considered safe when indicated, but it is essential to differentiate IIH from other pregnancy-related causes of increased ICP such as preeclampsia, eclampsia, and venous sinus thrombosis.

Management of Idiopathic intracranial hypertension in pregnancy :

Idiopathic intracranial hypertension (IIH) in pregnancy, though not directly life-threatening, can lead to significant maternal and fetal morbidity if inadequately managed. The most serious maternal complication is vision loss, which may result from progressive papilledema leading to optic atrophy and irreversible damage; early defects often include an enlarged blind spot or peripheral field constriction, with up to one-quarter of untreated patients developing permanent impairment. Persistent, severe headache is another disabling manifestation, and in pregnancy may be misattributed to preeclampsia, delaying targeted treatment.

Management of idiopathic intracranial hypertension (IIH) in pregnancy centers on two primary goals: preservation of maternal vision and symptom control, while ensuring fetal safety through the gestational period. Acetazolamide is the mainstay medical therapy for IIH, acting via reduction in cerebrospinal fluid production. Despite its FDA classification as category C, multiple observational series including over 150 pregnancies show no significant increase in congenital malformations or spontaneous abortion rates when used even in the first trimester. A systematic review of 178 IIH-affected pregnancies found acetazolamide to be first-line in over 50 cases, often combined with controlled weight gain. Some guidelines suggest postponing acetazolamide until after 20 weeks gestation, particularly if initiated for the first time, balancing theoretical risks with the real danger of visual loss.

Serial lumbar puncture (LP) may serve as a temporizing option in pregnant patients who either decline medication or require immediate symptom relief. Though CSF regenerates rapidly, LP may disrupt the vicious cycle of venous sinus stenosis seen in IIH and sometimes affords sustained improvement. However, it is labor-intensive and can be physically uncomfortable, especially in late-term pregnancy. Surgical interventions such as CSF diversion (lumboperitoneal or ventriculoperitoneal shunts) and optic nerve sheath fenestration (ONSF) are reserved for severe or vision-threatening cases. Both have been performed successfully in pregnancy when conservative measures fail. ONSF offers the advantage of being amenable to local anesthesia and can directly relieve papilledema, but carries risks including optic nerve injury and procedural failure. Endovascular stenting for transverse sinus stenosis shows promise outside pregnancy but is generally avoided in gestation due to prenatal radiation exposure and requirement for antiplatelet therapy.

IIH itself is not an indication for cesarean delivery. Studies show that neither vaginal delivery nor neuraxial anesthesia (spinal or epidural) adversely impact neurologic or visual outcomes even with in-situ shunts and epidurals can be safely administered, though caution is advised in shunted patients. When vision is threatened, a shortened second stage of labor may be prudent to avoid prolonged Valsalva-induced intracranial pressure rises. In our case, a general anesthesia approach was chosen due to uncertain lower uterine segment anatomy and to minimize intracranial fluctuations, aligning with reported safe general anesthesia use in IIH during cesarean section.

CONCLUSION :

This case highlights the importance of a multidisciplinary approach in managing pregnancy complicated by raised intracranial tension, particularly when associated with papilledema. Early detection, close monitoring, and coordinated care involving obstetrics, neurology, ophthalmology, and nephrology were pivotal in optimizing both maternal and fetal outcomes. The safe use of acetazolamide and sodium bicarbonate during pregnancy, under specialist guidance, contributed to neurological stability without compromising fetal well-being. Timely intervention with cesarean delivery at term allowed for a favorable perinatal outcome. This case underscores the need for individualized care plans in rare but high risk neurological complications during pregnancy.

REFERENCES:

1. **Lee AG, Pless M, Falardeau J, Capozzoli T, Wall M, Kardon RH.** *The use of acetazolamide in idiopathic intracranial hypertension during pregnancy. American Journal of Ophthalmology.* 2005;139(5):855–859.
2. **Falardeau J, Lobb BM, Golden S, Maxfield SD, Tanne E.** *The use of acetazolamide during pregnancy in intracranial hypertension patients. Journal of Neuro-Ophthalmology.* 2013;33(1):9–12
3. **Friedman DI, Jacobs DA, McDermott MP, et al.** *Managing IIH in pregnancy. Neurology* / clinical guidelines (via PMC).
4. **Huna-Baron R, Kupersmith MJ.** *Idiopathic intracranial hypertension and pregnancy. Journal of Neurology.* 2002;249(8):1078–1081.
5. **Case series and narrative review—IIH during pregnancy.** *Brazilian Journal of Anesthesiology (SciELO).*
6. **Byth LA, Lust K, Jeffree RL, Paine M, Voldanova L, Craven A-M.** *Management of idiopathic intracranial hypertension in pregnancy* (2022).
7. **Scott C, Kaliaperumal C.** *Idiopathic intracranial hypertension and pregnancy: a comprehensive review of management* (2022).