Other full case

Recurrent spontaneous intracranial hypotension in early pregnancy

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Summary

Spontaneous intracranial hypotension (SIH) is an uncommon condition characterised by postural headache secondary to low cerebrospinal fluid pressure. Here we present a case of recurrence of SIH in early pregnancy in a 26-year-old woman. She first presented at the age of 21 years at 15 weeks' gestation with a history of headache, nausea, vomiting, neck stiffness and photophobia. Findings from a MRI brain scan led to a diagnosis of SIH. She was treated with autologous epidural blood patching and remained asymptomatic until her second pregnancy 5 years later, when she re-presented at 16 weeks' gestation with similar symptoms. She was again diagnosed with SIH and required a repeat treatment of autologous epidural blood patching. She subsequently remained symptom free and delivered a healthy boy at term.

BACKGROUND

Spontaneous intracranial hypotension (SIH) is an uncommon condition associated with low cerebrospinal fluid (CSF) pressure. It was first described by Schaltenbrand in 1938 who described it as 'spontaneous aliquorrhoea'.¹ It is typically characterised by a postural headache, which is more severe in the upright position and relieved by lying recumbent.² Other features include photophobia, nausea, vomiting, anorexia, dizziness, neck stiffness, neck pain, tinnitus, vertigo, diplopia and blurred vision.^{1–8} The underlying pathogenesis is believed to be secondary to a dural tear with resultant spontaneous CSF leakage.^{2,3,9} Neuroimaging findings range from normal to subdural fluid collections such as haematomas and hygromas, dural thickening, pachymeningeal enhancement post gadolinium contrast, caudal cerebellar tonsillar and brainstem displacement and narrowing of the ambient cistern.^{2,10–12}

Typical findings on lumbar puncture include low or immeasurable CSF opening pressure. CSF examination itself is often normal although increased levels of CSF protein, erythrocytes and lymphocytes, as well as xanthochromia, have been reported.¹³ CT cisternography is useful in illustrating a dural tear. Other imaging techniques include CT myelography and spinal MRI myelography.

SIH is very rarely described in pregnancy. Furthermore, recurrence has not previously been reported in this setting making this an interesting case.

CASE PRESENTATION

A 21-year-old lady, gravida one para zero, presented at 15 weeks' gestation with a 1-week history of persistent nausea, vomiting, neck stiffness and photophobia. She also reported a 6-week history of intermittent frontal headache. The headache was dull in nature, occurred daily and constantly for the 4 days prior to admission, worsened on standing and was relieved on lying flat. She reported that symptoms of nausea, vomiting, neck stiffness and photo-

Figure 1 Time of flight MR venogram image illustrating a filling defect in the right transverse sinus.

phobia occurred in association with the headache for 1 week (International Classification of Headache Disorders II 7.2.3). There was no history of previous recurrent headaches, head/spinal trauma or intervention. It was also noted that there was no history of headaches or connective tissue disorders within her family. Her neurological examination, including funduscopy, was essentially normal. Her blood pressure on admission was 124/68. D-dimer level was within the normal range. A diagnosis of hyperemesis gravidarum was made but, due to the prominent headache, neuroimaging was performed. The MR venogram suggested

Figure 2 Time of flight MR venogram sagittal image illustrating a filling defect in the proximal right transverse sinus.

thrombosis of the right transverse venous sinus (figures 1 and 2). Thus, intravenous heparin was started. However, her headaches remained intractable and a neurological consultation was requested. A MRI brain scan demonstrated cerebellar tonsillar descent with ectopia and flattening of the pontine profile (figure 3). Gadolinium was not administered due to her early stage of pregnancy so presence of pachymeningeal enhancement could not be determined. The previous suggestion of venous sinus thrombosis was then believed to be an artefact after further opinions were obtained. Due to the prominent orthostatic component of her headache, consistent with a low CSF pressure, a lumbar puncture was performed. The opening pressure was <2 cm H_2O with sluggish and minimal CSF flow. She was diagnosed with SIH. The heparin was discontinued and she was treated with two autologous epidural blood patches, 72 h apart, which alleviated her symptoms. At follow-up 4 weeks later she remained asymptomatic and at term delivered a healthy boy via spontaneous vaginal delivery. No further headache occurred until she became pregnant 5 years later.

The second presentation of this, then 26-year-old, lady, gravida two para one, was at 16 weeks' gestation. She presented with a 10-day history of frontal headache with associated nausea and photophobia. The headache radiated towards the posterior aspect of the neck, occurred intermittently on a daily basis, worsened on standing and towards evening time and improved on lying recumbent. The headache became progressively more severe in the week prior to admission. There was no associated vomiting (International Classification of Headache Disorders II 7.2.3). There was no previous history of head or spinal trauma. Examination, including funduscopy, was unremarkable. Her blood pressure on admission was 116/64. Her D-dimer level was within the normal range.

Figure 3 T1 weighted MRI sagittal image illustrating cerebellar tonsillar ectopia.

She was initially treated with 3500 units of subcutaneous tinzaparin once a day to treat the possibility of cerebral venous sinus thrombosis while conservative measures, including bed rest, intravenous fluids, oral analgesics and increased caffeine intake, were used to treat the possibility of intracranial hypotension.

A MRI brain scan was performed, which revealed no abnormalities. In addition, normal venous sinuses were visualised on MR venography (figure 4). A MRI brain scan with gadolinium enhancement was not performed due to her early stage of pregnancy. A lumbar puncture was performed revealing a low opening pressure of 4 cm H_2O . She was again diagnosed with SIH. She failed to improve on conservative management so an autologous epidural blood patch procedure was performed with injection of 25 ml of autologous blood into the epidural space between the second and third lumbar vertebrae. This provided symptomatic and complete relief of headache.

OUTCOME AND FOLLOW-UP

At subsequent follow-up 4 weeks after discharge she remained asymptomatic. At term she delivered a healthy boy. There has been no recurrence of headache or other symptoms since delivery.

DISCUSSION

Headache, nausea and vomiting are common first trimester symptoms and may relate variably to hyperemesis gravidarum or to migraine, which can present de novo in pregnancy. The symptoms may also less commonly relate to SIH, as in this case.

A MEDLINE search of the literature performed from January 1966 to April 2010 using the search terms 'spontaneous intracranial hypotension', 'pregnancy' and 'headache' revealed 13 cases, 6 of which were relevant, in the

Figure 4 Time of flight MR venogram illustrating normal venous sinuses.

literature.^{14–19} A similar search with the addition of ‘recurrence’ revealed no cases. To the authors’ knowledge, this is the first case report in the literature reporting a recurrence of SIH in pregnancy.

SIH is poorly recognised and possibly under diagnosed. The present case suggests it may be specifically related to early pregnancy in some patients. Its recurrence in her second pregnancy with absence of symptoms outside of pregnancy would suggest this. A previous case report in the literature would also support this, in which the patient remained symptom free in the postpartum follow-up period.¹⁴ This may be a coincidence or there may be an underlying mechanism predisposing to SIH in pregnancy. One possibility is that physiological changes in intracranial pressure and CSF volume during pregnancy could predispose to this condition. However, the precise nature of the relationship is yet unclear.

Although our patient did not have the characteristic MRI features on repeat presentation, the clinical picture and her previous history, as well as her response to treatment, were all highly suggestive of a diagnosis of SIH. Indeed, atypical cases have previously been reported in the literature, including patients with absence of headache, absence of pachymeningeal enhancement post gadolinium enhanced MRI and normal CSF pressure.^{5 20–22} MRI with gadolinium contrast was not used due to concern about possible adverse effects on the neonate. CT myelography, which is normally used to locate the site of CSF leakage, was also not used due to concerns regarding its safety during pregnancy. This limits the ability to diagnose this condition in pregnancy as accurately as in the non-pregnancy state.

Of note, intracranial venous sinus thrombosis is a feared but rare complication of pregnancy. Non-visualisation or apparent obliteration of the transverse sinus does not necessarily mean thrombosis, but may occur as part of a haemodynamic consequence of low CSF pressure and downward

descent of posterior fossa structures. CSF analysis is crucial to the accurate classification of intractable headache in this setting. A subnormal CSF pressure virtually out rules significant intracranial sinus thrombosis and provides support for SIH, which remains predominantly a clinical diagnosis. It is worth highlighting to anaesthetic colleagues that epidural blood patching remains the current definitive treatment for SIH and repeat treatment may be necessary in cases of non-resolving symptoms.

Further study is required to accurately elucidate the possible association between SIH and its occurrence in pregnancy.

Learning points

- ▶ SIH is an uncommon cause of headache. It is poorly recognised and, therefore, possibly under diagnosed. However, it is important to consider it as a part of the differential diagnosis of headache particularly when occurring with associated symptoms such as nausea, vomiting and so forth.
- ▶ There may be an underlying mechanism predisposing to this condition specifically in pregnancy possibly relating to physiological changes in this state.
- ▶ Management consists of conservative measures such as bed rest, intravenous fluids, oral analgesia and increased caffeine intake. Autologous epidural blood patching is an alternative treatment if conservative measures fail to provide symptomatic relief.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Schaltenbrand G.** Neuere anschauungen zur pathophysiologie der liquorzirkulation. *Zentralbl Neurochir* 1938;**3**:290–300.
2. **Mokri B.** Headaches caused by decreased intracranial pressure: diagnosis and management. *Curr Opin Neurol* 2003;**16**:319–26.
3. **Rando TA, Fishman RA.** Spontaneous intracranial hypotension: report of two cases and review of the literature. *Neurology* 1992;**42**:481–7.
4. **Mokri B, Piegras DG, Miller GM.** Syndrome of orthostatic headaches and diffuse pachymeningeal gadolinium enhancement. *Mayo Clin Proc* 1997;**72**:400–13.
5. **Mokri B.** Spontaneous cerebrospinal fluid leaks: from intracranial hypotension to cerebrospinal fluid hypovolemia—evolution of a concept. *Mayo Clin Proc* 1999;**74**:1113–23.
6. **Mokri B, Posner JB.** Spontaneous intracranial hypotension: the broadening clinical and imaging spectrum of CSF leaks. *Neurology* 2000;**55**:1771–2.
7. **Brightbill TC, Goodwin RS, Ford RG.** Magnetic resonance imaging of intracranial hypotension syndrome with pathophysiological correlation. *Headache* 2000;**40**:292–9.
8. **Capobianco DJ, Kuczer FJ Jr.** Case report: primary intracranial hypotension. *Mil Med* 1990;**155**:64–6.
9. **Chung SJ, Kim JS, Lee MC.** Syndrome of cerebral spinal fluid hypovolemia: clinical and imaging features and outcome. *Neurology* 2000;**55**:1321–7.
10. **de Noronha RJ, Sharrack B, Hadjivassiliou M, et al.** Subdural haematoma: a potentially serious consequence of spontaneous intracranial hypotension. *J Neurol Neurosurg Psychiatr* 2003;**74**:752–5.
11. **Fishman RA, Dillon WP.** Dural enhancement and cerebral displacement secondary to intracranial hypotension. *Neurology* 1993;**43**:609–11.
12. **Pannullo SC, Reich JB, Krol G, et al.** MRI changes in intracranial hypotension. *Neurology* 1993;**43**:919–26.
13. **Ferrante E, Savino A, Sances G, et al.** Spontaneous intracranial hypotension syndrome: report of twelve cases. *Headache* 2004;**44**:615–22.
14. **Singh T, Schroeder F, Pereira A, et al.** Antenatal blood patch in a pregnant woman with spontaneous intracranial hypotension. *Int J Obstet Anesth* 2009;**18**:165–8.

15. **Jungmann V**, Werner R, Bergmann J, *et al.* [Postpartum cerebral venous sinus thrombosis after epidural anaesthesia]. *Anaesthesist* 2009;**58**:268–72.
16. **Albayram S**, Tuzgen S, Gbndpz A, *et al.* Spontaneous intracranial hypotension after labor without spinal intervention. *Eur J Neurol* 2008;**15**:91–3.
17. **Bel I**, Moreno LA, Gomar C. Epidural dextran-40 and paramethasone injection for treatment of spontaneous intracranial hypotension. *Can J Anaesth* 2006;**53**:591–4.
18. **Asakura H**, Hayashi Z, Seto M, *et al.* Spontaneous intracranial hypotension during pregnancy. *Obstet Gynecol* 2001;**97**:804–5.
19. **Carrero EJ**, Agustó M, Fábregas N, *et al.* Unilateral trigeminal and facial nerve palsies associated with epidural analgesia in labour. *Can J Anaesth* 1998;**45**:893–7.
20. **Kong DS**, Park K, Nam DH, *et al.* Atypical spontaneous intracranial hypotension (SIH) with nonorthostatic headache. *Headache* 2007;**47**:199–203.
21. **Mokri B**, Atkinson JL, Dodick DW, *et al.* Absent pachymeningeal gadolinium enhancement on cranial MRI despite symptomatic CSF leak. *Neurology* 1999;**53**:402–4.
22. **Mokri B**, Hunter SF, Atkinson JL, *et al.* Orthostatic headaches caused by CSF leak but with normal CSF pressures. *Neurology* 1998;**51**:786–90.

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