Beware of delayed severe brain swelling after intracerebral haematoma in HELLP syndrome

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Abstract

Haemolysis, elevated liver enzymes and low platelets (HELLP) syndrome is a condition associated with increased risk of adverse outcomes during pregnancy and peripartum, including neurological complications. We report the third case in the world literature of delayed brain swelling following cerebral haemorrhage as a complication of HELLP syndrome. A 36-year-old woman in labour developed HELLP, which was complicated with intracerebral haematoma. This was evacuated, but motor impairment persisted after surgery and unfortunately the patient died unexpectedly during the 11th post-operative day. Computer tomographic brain scans documented diffuse cerebral swelling, which we think may have been caused by cerebral vasospasm. Cerebral vasospasm should always be considered when managing patients who suffered from stroke complicating HELLP syndrome. Close monitoring is advised even in later stages of recovery.

Keywords

Cerebral swelling, HELLP syndrome, intracerebral haemorrhage, cerebral vasospasm

Introduction

The association between haemolysis, thrombocytopenia and liver dysfunction (HELLP [haemolysis, elevated liver enzymes and low platelets] syndrome) has been recognized more than five decades ago as a life-threatening complication of preeclampsia/eclampsia.^{1–3} The cerebral manifestations of HELLP syndrome are associated with significant adverse maternal outcome, and there has been scarce reporting in literature regarding intracerebral haemorrhage (ICH) in HELLP.^{2,4–6} We present a rare case of delayed fatal brain swelling, which followed an ICH complicating HELLP syndrome.

Case report

A 36-year-old primigravida was admitted in spontaneous labour to the Midwives Unit at 40 + 4 weeks gestation. Her pregnancy had been uncomplicated, and her past medical history was unremarkable. Her blood pressure (BP) on admission was 128/90 (booking BP 110/60). Three and a half hours after admission, at 08:00 hours, while her cervix was 6 cm dilated, she started complaining of headache and developed numbness in the left hemisoma and slurred speech. Physical examination documented a BP of 186/96, ankle oedema, left-sided weakness, left sensory inattention and left homonymous hemianopia. At that stage her cervix was fully dilated and she had a Neville Barnes forceps delivery at 09:20 with local anaesthesia to the perineum. This was uneventful and the newborn child was healthy. A computed tomographic (CT) brain scan taken soon after the delivery documented an ICH in the superior right centrum semiovale and extending into the lateral ventricles (Figure 1). She underwent a craniotomy with evacuation of haematoma and insertion of external ventricular drain (EVD). Intraoperatively, no abnormal vessels were identified.

Biochemical findings obtained after surgery showed thrombocytopenia (44,000/ μ L), anaemia (Hb 7.0 g/dL), deranged serum liver enzymes levels (aspartate aminotransferase 640 U/L, alanine aminotransferase 794 U/L), elevated serum lactate dehydrogenase (1295 U/L) and elevated serum total bilirubin (63 μ mol/L). The peripheral blood film was negative for haemolysis. A diagnosis of partial HELLP syndrome was made.^{7,8}

Over the following days physical examination showed left-sided dense hemiparesis associated with left upgoing plantar and left sixth

nerve palsy. She continued complaining of headache and remained hypertensive despite labetalol infusion (BP 168/90), while the Glasgow Coma Score (GCS) remained 15/15.

Despite continued cerebrospinal fluid (CSF) drainage her headache dramatically worsened on day 10, when it became associated with dysconjugate eye movements. There was some concern that the EVD may have been intermittently blocking. Despite the CSF examination showing no evidence of infection, it was decided to remove her EVD and insert a new one. Postoperatively she did not wake from anaesthesia and developed an abnormal breathing pattern. A CT brain scan showed that the ventricular catheter was within the frontal horn of the left ventricle although slightly too far advanced with the tip within brain parenchyma. The basal cisterns were still open. The EVD was withdrawn slightly but continued to drain only intermittently, and an intracranial pressure (ICP) bolt was placed. At this time her BP was 174/97 despite treatment and her GCS was E1 Vt M3. In the early morning of the 11th postoperative day the recorded ICP measurements went up rapidly to over 90 mmHg despite the administration of mannitol, and her pupils became fixed and dilated. Another CT brain scan documented marked intracerebral oedema with diffuse loss of grey-white matter differentiation (Figure 2). Decompressive craniectomy was considered but was felt likely to be futile.

Laboratory findings at this stage were unremarkable apart from an increased platelet count of 523,000/ μ L. On the same day brainstem tests confirmed death.

The postmortem brain examination demonstrated significant herniation of the medial temporal lobes and the cerebellar tonsils consistent with a final diagnosis of severe cerebral swelling. No definite underlying abnormality of vascular or other nature was identified within nor in the vicinity of the residual right fronto-parietal haematoma. No other areas of cerebral infarction were identified, and the

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Figure 1. Preoperative axial computed tomographic (CT) brain scan showing haematoma in the right centrum semiovale (left) and reconstructed coronal CT brain scan showing haemorrhage in the right parietal region extending into lateral ventricles and ventriculomegaly and evidence of midline shift (right).

Figure 2. Axial computed tomographic (CT) brain scans immediately following the revision of cerebrospinal fluid (CSF) drainage device (left) and 12 hours later (right) at the level of the pineal gland. There has been marked cerebral swelling with effacement of the lateral ventricles. The external ventricular drain is seen in the right frontal horn of the lateral ventricle.

macroscopic dissection of the Circle of Willis revealed no abnormalities.

Discussion

HELLP syndrome is a clinical condition attributed to a patient who develops haemolysis, elevated liver enzymes and low platelets in a context of pre-eclampsia/eclampsia.^{1,3,8,9}

It has been estimated that HELLP affects from 10% to 20% of patients with severe pre-eclampsia, and its mortality varies from 0% to 24%.¹ While in 30% of cases HELLP is diagnosed only in the post-natal period,² in the vast majority it occurs in the third trimester (27–37 weeks). Its aetiology remains elusive and yet incompletely understood.¹

Our patient suffered from an ICH as a complication of HELLP syndrome. While antenatally she was essentially well, the pre-eclampsia and HELLP presented in a fairly severe form and was clinically

diagnosed only during the intrapartum period. Although ICH is the most common among the neurological consequences of HELLP in postpartum period, a non-traumatic intracerebral bleeding is usually associated with vascular abnormalities such as arterial-venous malformations or neoplasms.^{7,10–12}

Nonetheless, the pathological examination of the blood clot evacuated during the original craniotomy did not demonstrate any vascular abnormalities or tumours, and the final neuropathological examination did not reveal a definite cause for the spontaneous ICH.

Sibai *et al.*¹³ recognized four cases of brain oedema in a prospective study involving 442 HELLP syndrome-complicated pregnancies. To the best of our knowledge, only two cases of delayed diffuse brain swelling complicating ICH in HELLP syndrome have been reported in literature so far.^{11,14} In these cases, an unexpected diffuse cerebral swelling occurred, and was attributed to a delayed cerebral vasospasm.

It is known that cerebral vasospasm can occur at up to 11 days after the onset of HELLP syndrome.¹¹ It has been hypothesized that pre-eclampsia itself reflects a subtype of hypertensive encephalopathy^{1,11,15} in which the forced vasodilatation and subsequent passive overdistention of cerebral arterioles leads to vasogenic oedema.^{5,16} Nonetheless, hypertensive encephalopathy could also lead to a prolonged reactive vasospasm with subsequent cytotoxic oedema and cerebral infarction.

Immediate delivery still remains the optimal treatment for HELLP syndrome diagnosed antenatally.^{1,2}

Control of stroke-range hypertension (over 170/110) and prevention of seizures are advised. Red cells and platelet transfusion is necessary in the more severe cases.¹

In this case we suspect that the cerebral swelling was not related to the original haemorrhage nor to the surgical trauma.^{11,17} We postulate that cerebral vasospasm occurred in the context of hypertensive encephalopathy related to the pre-eclamptic state. We recommend that the possibility of late cerebral swelling should be borne in mind in these cases. It is not clear whether such poor outcome could always be avoided but it may be worthwhile estimating cerebral blood flow with transcranial Doppler measurements or CT perfusion studies. Should there be a trend towards worsening perfusion one might consider early decompressive craniectomy before herniation syndromes occur. There is now reasonably good evidence that this treatment modality is effective in malignant middle cerebral artery strokes¹⁸ and prospective studies are underway in traumatic brain injury.¹⁹

Conclusion

We have presented an unfortunate case of death following delayed brain swelling as a complication of HELLP syndrome. The prognosis of HELLP syndrome remains severe. Early detection, experienced management in an intensive care environment associated with extended neuroradiological examination are desirable. Careful monitoring is recommended even in the later phases of recovery, and delayed vasospasm with haemodynamic alterations should always be considered. An improved understanding of the disease process may allow earlier recognition and improved outcome of this often fatal condition.

Declarations of Conflicting Interests

The authors declare that they have no competing interest.

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Ethical approval

No ethical approval was required for this work because the paper is a purely observational report, with no elements of intervention, and data were collected and thoroughly anonymized in an established educational setting.

Guarantor

AS.

Contributorship

GG and PAB conceived the case report. GG researched literature, collected data and wrote the first draft of the manuscript. EKL, PAB and AS reviewed the manuscript and approved its final version, which was written by GG. GG and PAB edited the manuscript revisions.

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