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Cerebral venous sinus thrombosis in pregnancy: a case report

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We report a case of peripartum cerebral venous sinus thrombosis in a 33-year-old Chinese multiparous woman with 38 weeks of gestation and uneventful antenatal history who presented with a 3-day history of progressively worsening generalised headache. The woman later developed recurrent convulsions. After joint consultation with the neurologist, neurosurgeon, and anaesthetist, emergency Caesarean section under general anaesthesia was performed with perioperative anticonvulsant cover and postoperative anticoagulation therapy. A high index of clinical suspicion and multidisciplinary discussion are important for its timely management.

Keywords: Headache; Pregnancy; Seizure; Sinus thrombosis, intracranial

Case presentation

In August 2014, a 33-year-old Chinese multiparous woman with 38 weeks of gestation and uneventful antenatal history presented with a 3-day history of progressively worsening generalised headache. She reported no history of head injury, convulsions, or other symptoms suggestive of pre-eclampsia. Her blood pressure was 126/76 mm Hg and she had no albuminuria. Neurological examination results were unremarkable. Results of urgent blood tests for complete blood count, serum electrolytes, liver enzymes, urate and coagulation profile were all normal. The cardiotocogram was normal.

Urgent non-contrast computed tomography (CT) of the brain showed hyperdensities in the superior sagittal sinus and cortical vein (Figure 1). There was no midline shift or space-occupying lesion. The diagnosis of cerebral venous sinus thrombosis (CVST) was suspected. The on-call neurologist, neurosurgeon, and radiologist were consulted. Urgent CT venogram revealed a venous thrombosis involving superior sagittal, right transverse, and right sigmoid sinuses extending to the origin of right internal jugular vein (Figure 2). The woman later developed two episodes of generalised convulsions, and intravenous levetiracetam was given as anticonvulsant. Neurological examination results were normal. The fetal heart tracing was all along reactive. After joint consultation with the neurologist, neurosurgeon, and anaesthetist, emergency Caesarean section under general anaesthesia was performed with perioperative anticonvulsant cover and postoperative anticoagulation therapy. The operation was uncomplicated, with blood loss of 400 mL. A baby girl weighing 4.04 kg with good Apgar scores was delivered.

The woman was transferred to the intensive care unit for further management. She was prescribed subcutaneous low-molecular-weight heparin (LMWH) after haemostasis was ascertained. She recovered well and had no more convulsions or neurological deficits. On postoperative day 5, she requested to be managed in the private sector. A referral letter was issued about the peripartum events and the increased risk of venous thromboembolism in future pregnancy. She planned to use male condoms for contraception.

Discussion

CVST is rare and characterised by thrombosis of the dural sinuses and cerebral veins, causing venous congestion, cerebral oedema, haemorrhagic venous infarctions, and neuronal damages. In developed countries, its incidence is 11.6 per 100 000 pregnancies[1]. It is more common in the third trimester, and >75% of cases occur postpartum[2]. The increased risk of CVST in pregnancy is related to hypercoagulability and is higher in those with hypertension, advanced maternal age, Caesarean delivery, infections, or dehydration exacerbated by excess vomiting[3].

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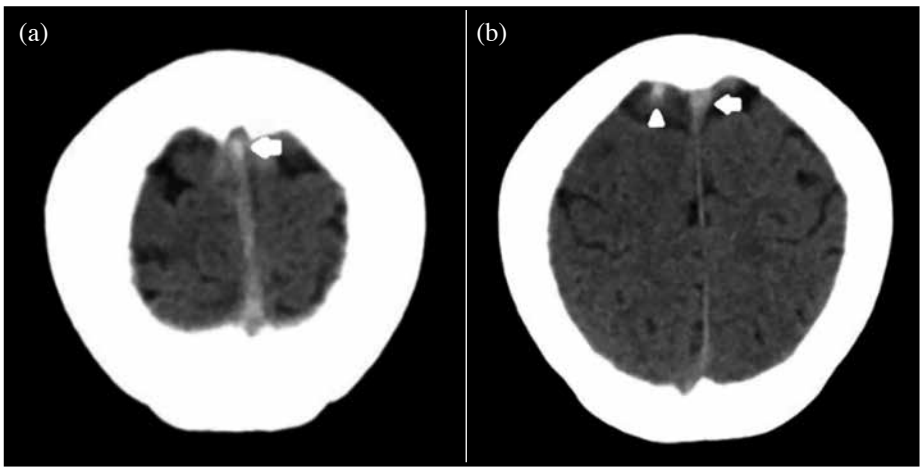

Figure 1. Non-contrast computed tomography of the brain showing hyperdensities at the superior sagittal sinus (arrows) and the cortical vein (arrowhead) suspicious of cerebral venous sinus thrombosis.

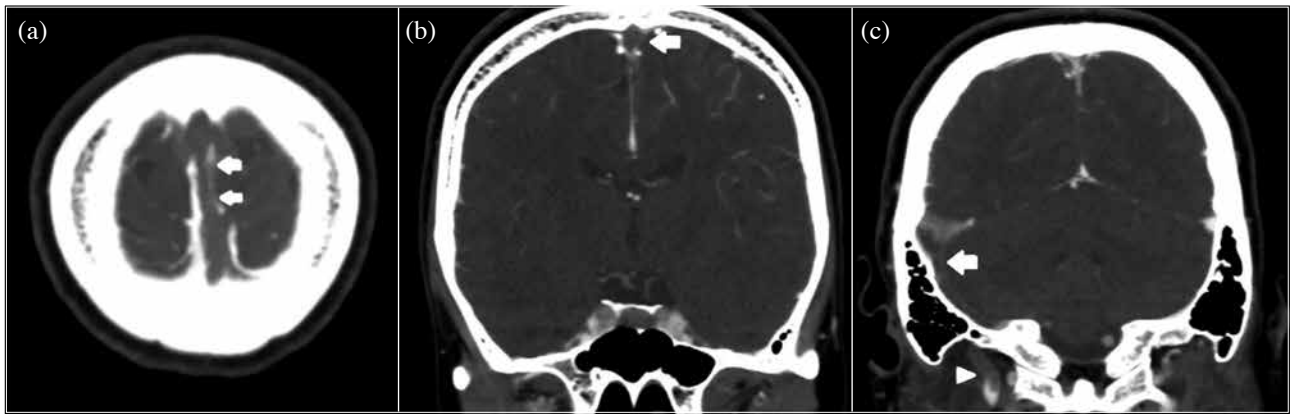

Figure 2. Computed tomographic venography showing filling defects at the (a and b) superior sagittal sinus, with the 'empty delta sign' (arrow) and (c) right transverse and right sigmoid (arrow) sinuses extending to the origin of the right internal jugular vein (arrowhead). The diagnosis of cerebral venous sinus thrombosis is confirmed.

The diagnosis of CVST requires a high degree of clinical suspicion owing to its rarity and variable clinical presentation. Headache is the most frequent symptom in CVST, but headache is a common complaint in pregnancy and postpartum period, and >90% of headaches are primary and benign and self-limiting without neurological sequalae[4]. Most patients with CVST present with a progressively diffuse headache, and 10% of them have an abrupt severe headache (as known as thunderclap headache). Headache could be the only presenting feature, but 10% of patients with CVST have no headache at all[2]. It is important to note that symptoms of CVST vary and can fluctuate over time, and that neurological deficits do not follow arterial distribution. Specific neurological presentation depends on the extent and location of venous thrombosis and its resultant complications. Other symptoms include dizziness, nausea, lethargy, visual loss, diplopia, and papilloedema. In a meta-analysis of observational studies, pregnancy is a significant factor associated with early-onset seizure (odds ratio [OR]=2.054)[5]. 40% of patients with CVST have focal or generalised seizures, which should alert physicians the need for further neuroimaging.[2]

Non-contrast CT are often negative, but it may demonstrate hyperdense thrombosed cortical veins or dural sinuses in a third of patients in the acute phase.[6] Ischaemic infarcts often undergo haemorrhagic transformation secondary to venous congestion and hypertension[2]. Parenchymal abnormalities such as cerebral oedema and haemorrhages may be seen most conspicuously on magnetic resonance imaging of the brain. Both CT venography and magnetic resonance venography (MRV) have comparable sensitivity for diagnosing CVST[7]. Nonetheless, there are concerns of radiation risk and drug safety, in addition to limited availability. For CT of the brain, as fetus is outside the field of view, the scattered radiation to the fetus is

negligible, and thus medical physicist consultation is not mandatory[8]. Contrast magnetic resonance venography is less preferable in pregnant women[8], as gadolinium-based contrast is considered as category C (ie, there are adverse effects on fetus in animal reproduction studies, but there are no controlled studies in humans), whereas iodinated contrast is considered as category B (ie, no adverse effects in animal reproductive studies, but there are no controlled studies in pregnant women). Non-contrast magnetic resonance venography using phase contrast or time-of-flight technique are feasible, but the results are sometimes inconclusive owing to artefacts.

Anticoagulation is the mainstay of treatment for CVST[7]. In a non-randomised study comparing patients receiving LMWH or unfractionated heparin, LMWH is associated with better functional independence after 6 months (adjusted OR=2.4) and fewer new intracerebral haemorrhages (adjusted OR=0.29)[9]. In a systemic review and meta-analysis, compared with unfractionated heparin, LMWH is recommended in the acute phase to reduce the mortality (OR=0.21)[10]. In the peripartum period, the optimal time to start anticoagulation requires balance between the thrombotic risk and bleeding risk during delivery and puerperium. The management for CVST in our patient with recurrent seizures was difficult. Discontinuation of anticoagulation is required before a scheduled delivery. Although seizures or epilepsy do not necessitate Caesarean delivery in general, the chance of recurrent seizure was high, and labour stress may increase the chance of convulsions compromising fetal well-being. Therefore, after multidisciplinary joint consultation, emergency Caesarean section was performed with perioperative anticonvulsant and postoperative anticoagulation when haemostasis was achieved.

Acute phase anticoagulation by LMWH, followed by oral anticoagulant for at least 3 to 12 months is recommended for patients with CVST[7]. The overall mortality rate of CVST was 2% to 10%, but the rate is significantly lower in pregnancy-associated cases[1,6]. According to the International Study on Cerebral Vein and Dural Sinus Thrombosis, most patients with CVST will have none (79%) to mild (8%) residual deficits after treatment[11]. Severe headache (14.1%), seizures (10.6%), and new thrombotic events (4.3%) are the most frequent complications during follow-up. Women with a history of CVST are advised against oestrogen-containing contraceptives. LMWH prophylaxis should be prescribed during subsequent pregnancies. Risks for recurrent CVST and non-cerebral venous thromboembolism in future pregnancies are reported to be 9 and 27 per 1000 pregnancies, respectively, compared with 0.116 and 1.72 per 1000 pregnancies in the general obstetric population[12]. Thrombophilia testing is recommended for women with thrombotic events in pregnancy and puerperium[13]. However regardless of thrombophilia abnormalities, they have a higher chance of adverse late obstetric events, including small-for-gestational age newborn, pre-eclampsia, eclampsia, HELLP syndrome (haemolysis, elevated liver enzymes, and a low platelet count), and placental abruption in their subsequent pregnancies, when compared with healthy women (19.2% to 24% vs 4%)[14,15]

Declaration

The authors have no conflict of interest to disclose.

Ethics approval

Ethics approval was obtained from New Territories West Cluster Research Ethics Committee (Ref: NTWC/REC/19097).

References

1. Lanska DJ, Kryscio RJ. Risk factors for peripartum and postpartum stroke and intracranial venous thrombosis. Stroke 2000;31:1274-82. Crossref
2. Edlow AG, Edlow BL, Edlow JA. Diagnosis of acute neurologic emergencies in pregnant and postpartum women. Emerg Med Clin North Am 2016;34:943-65. Crossref
3. James AH, Bushnell CD, Jamison MG, Myers ER. Incidence and risk factors for stroke in pregnancy and the puerperium. Obstet Gynecol 2005;106:509-16. Crossref
4. O'Neal MA. Headaches complicating pregnancy and the postpartum period. Pract Neurol 2017;17:191-202. Crossref
5. Li H, Cui L, Chen Z, Chen Y. Risk factors for early-onset seizures in patients with cerebral venous sinus thrombosis: a meta-analysis of observational studies. Seizure 2019;72:33-9. Crossref
6. Mortimer AM, Bradley MD, Likeman M, Stoodley NG, Renowden SA. Cranial neuroimaging in pregnancy and the post-partum period. Clin Radiol 2013;68:500-8. Crossref
7. Ferro JM, Bousser MG, Canhão P, et al. European Stroke Organization guideline for the diagnosis and treatment of cerebral venous thrombosis - endorsed by the European Academy of Neurology. Eur Stroke J 2017;2:195-221. Crossref

8. Tirada N, Dreizin D, Khati NJ, Akin EA, Zeman RK. Imaging pregnant and lactating patients. Radiographics 2015;35:1751-65. Crossref
9. Coutinho JM, Ferro JM, Canhão P, et al. Unfractionated or low-molecular weight heparin for the treatment of cerebral venous thrombosis. Stroke 2010;41:2575-80. Crossref
10. Al Rawahi B, Almegren M, Carrier M. The efficacy and safety of anticoagulation in cerebral vein thrombosis: a systematic review and meta-analysis. Thromb Res 2018;169:135-9. Crossref
11. Ferro JM, Canhão P, Stam J, Bousser MG, Barinagarrementeria F; ISCVT Investigators. Prognosis of cerebral vein and dural sinus thrombosis: results of the International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVT). Stroke 2004;35:664-70. Crossref
12. Aguiar de Sousa D, Canhão P, Ferro JM. Safety of pregnancy after cerebral venous thrombosis: systematic review update. J Neurol 2018;265:211-2. Crossref
13. Royal College of Obstetricians and Gynaecologists. ROCG Green-top Guideline No. 37b: thromboembolic disease in pregnancy and the puerperium: acute management. London: RCOG; 2015.
14. Abbattista M, Capecchi M, Martinelli I. Pregnancy after cerebral venous thrombosis. Thromb Res 2019;181(Suppl 1):S15-S18. Crossref
15. Martinelli I, Abbattista M, Somigliana E, Gianniello F, Peyvandi F. Pregnancy outcome after a first episode of cerebral vein thrombosis: reply. J Thromb Haemost 2017;15:1526. Crossref