

Acute cerebral venous sinus thrombosis presenting in papilloedema: a case report

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Abstract

Background: Papilloedema has several aetiologies, including brain tumours, central nervous system inflammation, cerebral venous thrombosis, cerebral venous sinus thrombosis (CVST), and idiopathic intracranial hypertension. CVST is a rare condition and a form of stroke with an incidence of 5:1,000,000/year.

Case presentation: A 23-year-old female developed impaired vision in her left eye, accompanied by headache, nausea, and vomiting, 5 weeks prior to hospitalization. Visual acuity in the left eye (OS) was 6/12, which improved to 6/9 on pinhole examination, and to 6/6 with S-0.50 and C-0.50 on the 180° axis. OS intraocular pressure (IOP) was 14 mmHg. Bilateral fundusoscopic examination revealed papilloedema, peripapillary venous tortuosity, and macular exudate. Both computerised tomography scan and magnetic resonance angiography examination detected superior sagittal sinus thrombosis.

Conclusion: Acute CVST is a rare condition. A good prognosis depends on early diagnosis and prompt treatment.

Keywords: neuro-ophthalmology, papilloedema, superior sagittal sinus thrombosis

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Manifestasi klinikal trombosis vena sinus serebral akut sebagai papiloedema: satu lapuran kes

Latarbelakang: Terdapat pelbagai penyebab papiloedema termasuklah ketumbuhan didalam otak, radang pada system saraf pusat, thrombosis vena sinus serebral (TVSS), dan hipertensi intracranial tanpa sebarang penyebab. TVSS adalah penyebab jarang jumpa dan merupakan salah satu jenis strok dengan insiden 5:1,000,000/setahun.

Persembahan kes: Seorang Wanita berumur 23 tahun mengalami pengurangan penglihatan mata kiri dan dituruti oleh sakit kepala, rasa mual dan muntah. Lima minggu sebelum dimasukkan ke hospital, ketajaman penglihatan mata kiri beliau adalah 6/12 dan ditambah baik dengan lubang pin kepada 6/9 seterusnya menjadi 6/6 dengan pembetulan refraktif sferikal -0.5 dan silendrikal -0.5 pada paksi 180 darjah. Tekanan mata kiri adalah 14 mmHg. Pemeriksaan funduskopi mendapati kehadiran papiloedema, vena periferak yang berliku-liku dan eksudat pada macular dikesan dikedua-dua fundus. Skan CT dan angiografi resonan magnetic mendapati terdapat thrombosis sinus sagittal pada bahagian atas.

Kesimpulan: TVTS akut adalah keadaan yang jarang berlaku. Prognosis yang baik bergantung kepada diagnose yang dibuat awal dan rawatan yang bertepatan pada masanya.

Kata kunci: neuroftalmologi, papiloedema, thrombosis sinus sagittal pada bahagian atas

Introduction

Papilloedema is optic disc swelling caused by elevated intracranial pressure (ICP).¹ In most cases, the swelling is bilateral and can occur over hours to weeks.² Papilloedema has several etiologies, including brain tumors, central nervous system inflammation, cerebral venous thrombosis, cerebral venous sinus thrombosis, and idiopathic intracranial hypertension.³ Headache is the most common cardinal feature in patients with increased ICP. When accompanied by nausea and vomiting, they specifically point to intracranial hypertension as a cause. However, those symptoms may also be found in migraine.²

Cerebral venous sinus thrombosis (CVST) is a rare form of thrombosis typically affecting young adults with a mean age of 35 years. It is more commonly found in females compared to males (2.2:1) due to gender-specific risk factors.⁴ CVST is primarily found in any prothrombotic condition, namely oral contraceptive consumption, pregnancy, and the post-partum state. As many as 70–80% of cases are reported in women of childbearing age.⁵ CVST has an incidence of 2–5 cases per million individuals per year, although it might be underestimated.⁴ The superior

sagittal and the transverse sinuses are the most commonly affected sinuses (60% of patients), followed by the internal jugular and cortical veins (20%).⁶ This case report presents a rare CVST case in a young female adult with papilloedema.

Case presentation

On February 4, 2021, a 23-year-old female came to the hospital with a primary complaint of blurred vision in the left eye (OS) for 5 weeks. It was accompanied by a headache, nausea, and vomiting. The patient had no complaints about her right eye (OD). On her visit, the patient mentioned that her symptoms persisted and worsened. The patient had neither a history of systemic illnesses nor eye complaints. The patient had never worn spectacles or other vision correction tools, or undergone eye surgery. Ophthalmological examination on the OS revealed a visual acuity of 6/12, which improved into 6/9 with a pinhole examination, and into 6/6 with S-0.50 and C-0.50 on the 180° axis. The OD has normal visual acuity (6/6). Both optic discs were hyperaemic and had blurred margins, vessel obscuration, and tortuous veins (Fig. 1). The intraocular pressure (IOP) was 18 mmHg for the OD and 14 mmHg for the OS. The Ishihara test showed normal results for both eyes. The contrast sensitivity test gave 2.33 for the OD and 8.91 for the OS. Perimetry examination (Humphrey Field Analyzer, HFA) on the OD gave a fixation loss of 1/15 with a false positive error of 1%, a false negative error of 16%, and a visual field index (VFI) of 89%. For the OS, the fixation losses were 7/17 with a false positive error of 0%, a false negative error of 5%, and a VFI of 81% (Fig. 2). The HFA examination showed a specific papilloedema pattern (an enlarged blind spot), decreased mean deviation, and positive pattern standard deviation. These findings indicated a decreasing visual field sensitivity. The optical coherence tomography (OCT) analysis is shown in Figure 3. On the first day of visit (DOV), a marked disc swelling was presented by increased average retinal nerve fibre layer thickness, although it decreased in subsequent follow-up visits. However, the undefined cup volume resembled a subtle disc swelling condition.

Laboratory examination showed low haemoglobin levels (11/8 g/dl), high leucocyte count (12×10^3 cells/ μ L), normal thrombocyte count (321×10^3 cells/ μ L), high erythrocyte sedimentation rate (73 mm/1 hour), and high C-reactive protein (26 mg/dl). The patient's coagulation profile had normal results: 12 seconds of prothrombin time, 31.9 seconds of activated partial thromboplastin time, and 1.1 international normalised ratio. Computed tomography (CT) scan of the head with contrast detected a dural sinus thrombosis on the superior sagittal sinus, diffuse cerebral oedema, right optical nerve tortuosity, and retention cyst on the left maxillary sinus (Fig. 4). The patient was administered mecobalamin 500 mg tablets once daily and was referred to the neurosurgery division for a magnetic resonance angiography (MRA) with contrast examination. The MRA with

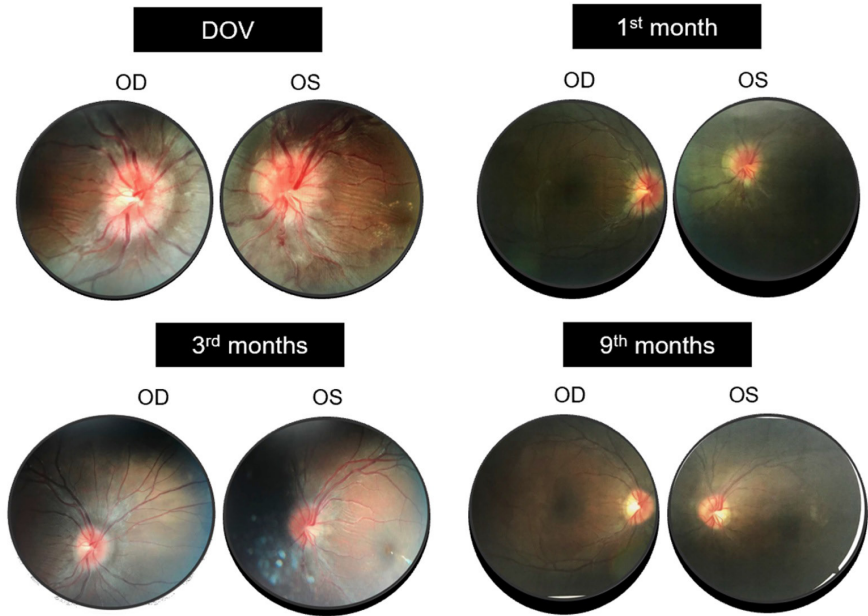


Fig. 1. Fundusoscopic examination on the first day of visit (DOV) and the first, third, and ninth months after treatment.

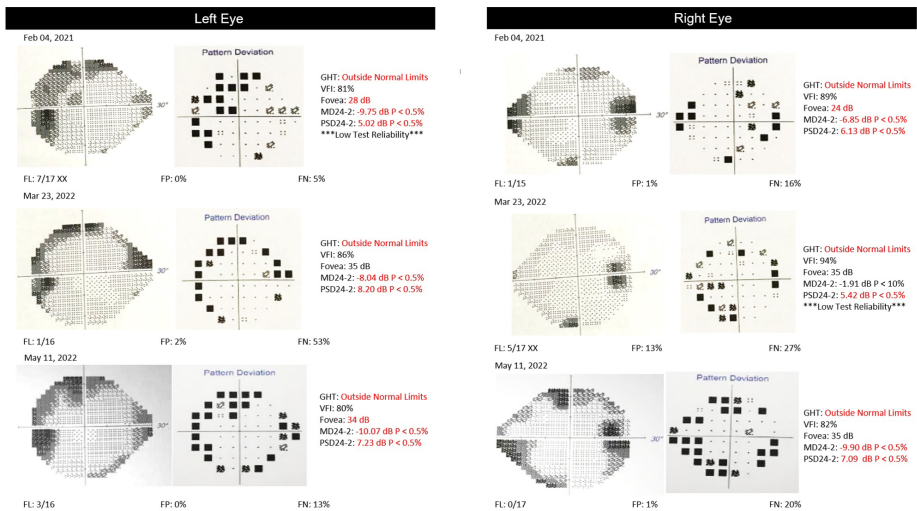


Fig. 2. Perimetry examination on the first day of visit (DOV) and the third and ninth months after treatment.

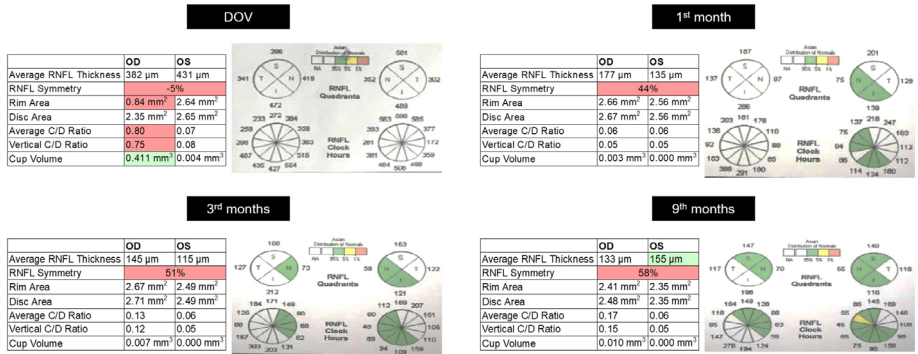


Fig. 3. OCT examination on the first day of visit (DOV) and the first, third, and ninth months after treatment.

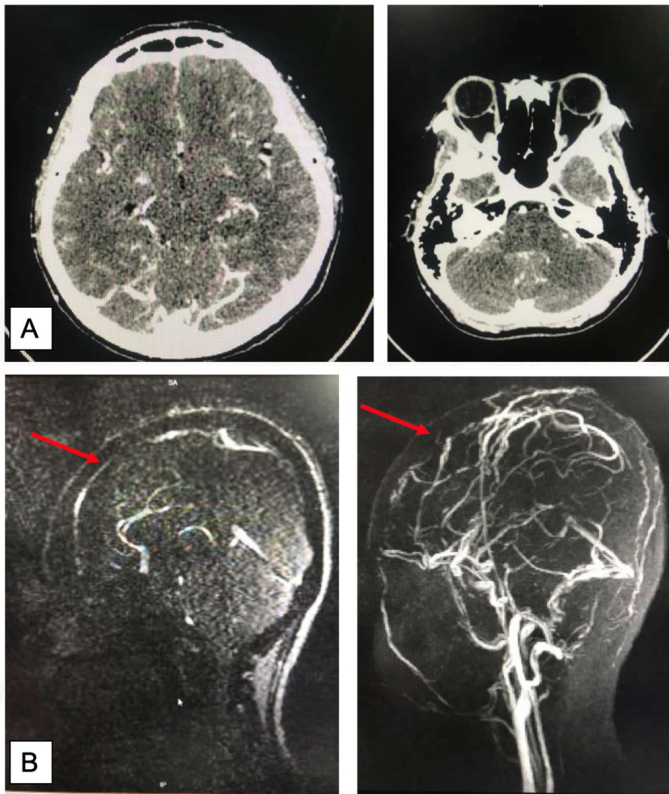


Fig. 4. (A) CT scan of the head with contrast revealed diffuse cerebral oedema and right optic nerve tortuosity. (B) The MRA with contrast shows thrombosis on the superior sagittal sinus (red arrow).

contrast analysis revealed a thrombosis on the superior sagittal sinus (Fig. 4). The neurosurgeon prescribed acetylsalicylic acid 80 mg tablets once daily, planned for another MRA evaluation test in the next 3 months, and referred the patient to the internal medicine department. The follow-up MRA with contrast examination showed a narrow superior sagittal sinus. The internist requested 2 additional examinations: an antinuclear antibody test and a rheumatoid arthritis/rheumatoid factor test. The results for both tests were negative. However, she started to have a painless ulcer on her right hallux. Therefore, the patient was referred to the surgical department, where she had a lower extremity ultrasonography examination, which detected an anterior tibial artery occlusion. The patient was diagnosed with gangrene, and amputation was suggested.

Discussion

This case report discusses a rare CVST disease in a 23-year-old female with bilateral papilloedema. Both the CT scan and MRA examination revealed superior sagittal sinus thrombosis. Aside from being diagnosed with CVST in the superior sagittal sinus, the patient also had peripheral arterial disease, which may suggest a similar pathophysiology. Conservative therapy without any thrombolytic or surgery was chosen for this patient. Thrombolysis should only be used in highly selected patients due to the risk of bleeding. The patient was given an anticoagulant (acetylsalicylic acid) to prevent new clot formation.

CVST is widely known as one of the primary causes of papilledema. In CVST, venous outflow obstruction leads to increased venous pressure, eventually causing venous stasis. The increasing venous pressure directly increases venular and capillary pressure or indirectly elevates ICP, causing headaches, seizures, focal neurological deficit, and papilloedema.⁷ An obstruction, such as thrombosis or compression on the CVS drainage, may cause high ICP. The obstruction commonly affects the superior sagittal and transverse (lateral) sinuses. Papilloedema occurs early, bilaterally, and symmetrically in CVST. Both children and adults are prone to aseptic thrombosis, most of which is found within the superior sagittal sinus. Patients may have an underlying coagulopathy (*e.g.*, factor V Leiden mutation, prothrombin gene 20210GA transition, activated protein C resistance, protein C, S, or antithrombin III deficiency, anticardiolipin antibodies, or hyperviscosity syndromes), systemic conditions (*e.g.*, renal disease, pregnancy, and cancer), predisposing medication use (*e.g.*, oral contraceptives), or systemic inflammatory or infectious diseases (*e.g.*, systemic lupus erythematosus, Behçet disease, trichinosis, or sarcoidosis).¹

The gold standard for diagnosing CVST is magnetic resonance imaging and magnetic resonance venography. They are more sensitive in recognising the presence of parenchymal oedema compared to CT scans. Acute thrombus (0 to 7

days) is hard to detect. It becomes easier to detect starting at week 2, where both T1- and T2-weighted images give hyperdense signals. In general, a non-contrast CT scan is the first test taken due to its speed and accessibility.⁸ A cord sign, a hyperattenuating curvilinear within a cortical vein in the presence of thrombosis, is a direct sign of CVST that may present for up to 2 weeks. Aside from a cord sign, a dense triangle sign may also be found. This sign is a hyperdense triangular shape in the superior sagittal sinus.⁵

A rare complication of CVST is a dural arteriovenous fistula, which may result from recurring papilledema (the estimated incidence rate is 1–3% based on cohort studies without systemic angiographic follow-up).⁸ The definitive treatment for CVST combines mechanical thrombectomy and thrombolysis. This combination resulted in 87% complete recanalization.⁹ This intervention is usually used in severe CVST and unresolved CVST after conservative anticoagulant or thrombolysis therapy alone.

Conclusion

The aetiologies of papilloedema include brain tumours, central nervous system inflammation, idiopathic intracranial hypertension, and CSVT. It is vital to recognize the presence of papilloedema and choose the appropriate supporting examinations to determine its cause. A good prognosis depends on early diagnosis and prompt treatment.

Declarations

Consent for publication

Written informed consent for the publication of the clinical data and images contained in this case report was obtained from the patient.

Competing interests

None to declare.

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References

1. Rigi M, Almarzouqi SJ, Morgan ML, et al. Papilledema: epidemiology, etiology, and clinical management. *Eye Brain*. 2015 Aug;17(7):47-57. <https://doi.org/10.2147/eb.s69174>
2. Overview and differential diagnosis of papilledema [Internet]. Massachusetts: UpToDate (US); [cited 2022 November 1]. Available from: <https://www.uptodate.com/contents/overview-and-differential-diagnosis-of-papilledema>
3. Papilledema [Internet]. Washington DC: American Academy of Ophthalmology (US); [cited 2022 November 1]. Available from: <https://eyewiki.aao.org/Papilledema>
4. Bousser MG, Ferro JM. Cerebral venous thrombosis: an update. *Lancet Neurol*. 2007 Feb;6(2):162-70. [https://doi.org/10.1016/s1474-4422\(07\)70029-7](https://doi.org/10.1016/s1474-4422(07)70029-7)
5. Tadi P, Behgam B, Baruffi S. Cerebral Venous Thrombosis. [Updated 2022 Jun 17]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK459315>
6. Maali L, Khan S, Qeadan F, et al. Cerebral venous thrombosis: continental disparities. *Neurol Sci*. 2017 Nov;38(11):1963-8. <https://doi.org/10.1007/s10072-017-3082-7>
7. Park, MG., Roh, J., Ahn, SH. et al. Papilledema and venous stasis in patients with cerebral venous and sinus thrombosis. *BMC Neurol* 23, 175 (2023). <https://doi.org/10.1186/s12883-023-03228-0>
8. Saposnik G, Barinagarrementeria F, Brown RD Jr, et al. Diagnosis and management of cerebral venous thrombosis: a statement for healthcare professionals from the American Heart Association/American Stroke Association. *Stroke*. 2011 Apr;42(4):1158-92. <https://doi.org/10.1161/str.0b013e31820a8364>
9. Li G, Zeng X, Hussain M, Meng R, et al. Safety and validity of mechanical thrombectomy and thrombolysis on severe cerebral venous sinus thrombosis. *Neurosurgery*. 2013 May 1;72(5):730-8. <https://doi.org/10.1227/NEU.0b013e318285c1d3>