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CASE REPORT

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Coexistence of cerebral venous thrombosis and dural arteriovenous fistula in an adolescent: A case report

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Key Clinical Message

Clinicians should consider central venous thrombosis (CVT) as a differential diagnosis in young adolescents with persistent headaches. It is essential to assess for concurrent CVT and dural arteriovenous fistula (DAVF), particularly in those with a history of CVT.

Abstract

Cerebral venous thrombosis (CVT) and dural arteriovenous fistula (DAVF) are uncommon vascular disorders with diverse clinical presentations. The coexistence of CVT and DAVF is a rare but important association that may impact the management and prognosis of affected patients. Prothrombotic conditions generally ranging from acquired to genetic, oral contraceptives, malignancy, puerperium, infection, and head injury are the common risk factors for cerebral venous thrombosis. Here, we present a case of 18 years males who developed recurrent cerebral thrombosis on the background of the presence of an arteriovenous fistula.

KEYWORDS

anti-coagulant, cerebral venous thrombosis, dural arteriovenous fistula, thrombosis

1 | INTRODUCTION

Cerebral venous thrombosis (CVT) is an uncommon disease of the venous system. It is usually undiagnosed due to its varied presentation ranging from new-onset headache, seizure, altered mental status, and focal neurological deficit.[1] It generally affects adults with a mean age of 35 years and mainly involves women.[2] Risk factors for cerebral venous thrombosis vary from genetics, infections, inflammation, and use of medications with hematological malignancy.[3] Long-term cerebral venous thrombosis can lead to visual deficit.[4] Dural arteriovenous fistula (DAVF) is another uncommon cerebrovascular disorder that typically manifests in middle-aged adults, with a higher occurrence among women. The occurrence of DAVF concomitant with CVT is less frequent. The question of whether DAVF is the underlying cause or a consequence of CVT, as well as the nature of DAVF lesions as congenital or acquired, continues to be a matter of controversy.[5] We reported a case of chronic cerebral venous thrombosis in a background of arteriovenous fistula with a visual deficit.

2 | CASE PRESENTATION

An 18-year-old male with a known case of cerebral venous sinus thrombosis for 1 year presented to our hospital with a history of vomiting and headache for 4 months and decreased vision in the right eye for the same duration. The patient reports experiencing multiple episodes of projectile vomiting without bile or blood, along with a persistent headache for the past 1 month. The headache is described as dull and non-pulsatile, without any seizure-like activity. Additionally, the patient has noticed decreased vision in the right eye for the same duration, particularly in the peripheral vision. There is no history of double vision, difficulty in identifying colors, deviation of the angle of the mouth, hearing impairment, swallowing difficulties, or altered sensorium. The patient was under anticoagulation therapy for past events and also gave a history of warfarin free time for about 1 month's duration before the onset of the symptom.

On examination, he was well-oriented to person, place, and time with a Glasgow coma scale (GCS) score of 15/15 (E4V5M6). His pulse rate was 86 beats/min, blood pressure was 100/70 mmHg, body temperature was 98°F, respiratory rate was 16 breaths/min, and oxygen saturation was 95% in room air. Systemic examinations did not reveal any significant abnormalities. However, bilateral papilledema was observed during fundus examination.

Laboratory investigations showed hemoglobin 14.9 g/dL and hematocrit 46.6%. The total leukocyte count was 11,700/mm^3, neutrophils were 78%, and platelet count was 208,000/mm^3. The prothrombin time was 24.7 s, and the international normalized ratio (INR) was 2.5. The protein S activity was 64% with a functional assay of 64% whereas protein C activity in plasma was 90% with a functional assay of 90%. The antithrombin activity was 88% The D-dimer, VDRL, ANA, C-reactive protein, RA factor, and serology tests were negative. On cerebrospinal fluid (CSF) analysis, sugar was 70 mg%, protein was 76 mg%, ADA was 8.1 U/L, and LDH was 56 U/L.

A plain computed tomography (CT) scan of the brain showed bilateral hypodensity on the area of the transverse sinus (Figure 1). A plain magnetic resonance imaging (MRI) study of the head showed abnormal hypointense signal along the course of bilateral transverse venous sinuses including torcula heterophil on T2/FLAIR while magnetic resonance venography (MRV) showed heterogenous altered signal flow in bilateral transverse venous sinuses (Figure 2). Multiple collateral vessels were seen arising from bilateral transverse venous sinuses connecting to the different circulation. Cerebral digital subtraction angiography (DSA) showed retrograde leptomeningeal venous drainage along with multiple feeders without intervening nidus (Figure 3). Superior sagittal sinus, inferior sagittal sinus, straight sinus, bilateral transverse, and sigmoid sinus as well as visible cortical veins were grossly dilated suggestive of bilateral dural arteriovenous fistula with multiple collaterals. No embolization for the fistula was performed.

Patient was admitted to ward with close observation and managed conservatively. He received acetazolamide 250 mg four times a day, enoxaparin 60 mg twice daily for 5 days, and bridging therapy was done. The patient was prescribed warfarin 5 mg once daily. The hospital course remained uneventful. The patient's symptoms improved, and they were discharged after 8 days. Acetazolamide and warfarin were continued until the next follow-up appointment.

3 | DISCUSSION

Dural venous and cerebral venous thrombosis is less common and difficult to diagnose. With increasing clinical awareness along with the use of dynamic radiological investigation, there are increasing cases of venous thrombosis. It usually affects middle-aged patients with more predominance of females than males.[6] Risk factors range from reversible to irreversible ones. It includes hereditary protein C and protein S deficiency, dehydration, use of oral contraceptive pills, substance abuse, thrombophilias, head trauma, infections, connective tissue disorder, cancer, and many more.[6,7]

Clinical presentation of the CVT is non-specific so it is difficult to diagnose.[6] Variable clinical presentation

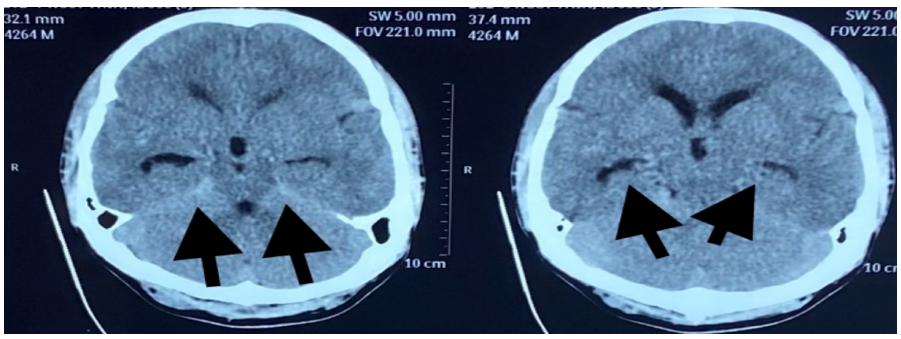

FIGURE 1 NCCT head showing hyperdensity on bilateral transverse sinus.

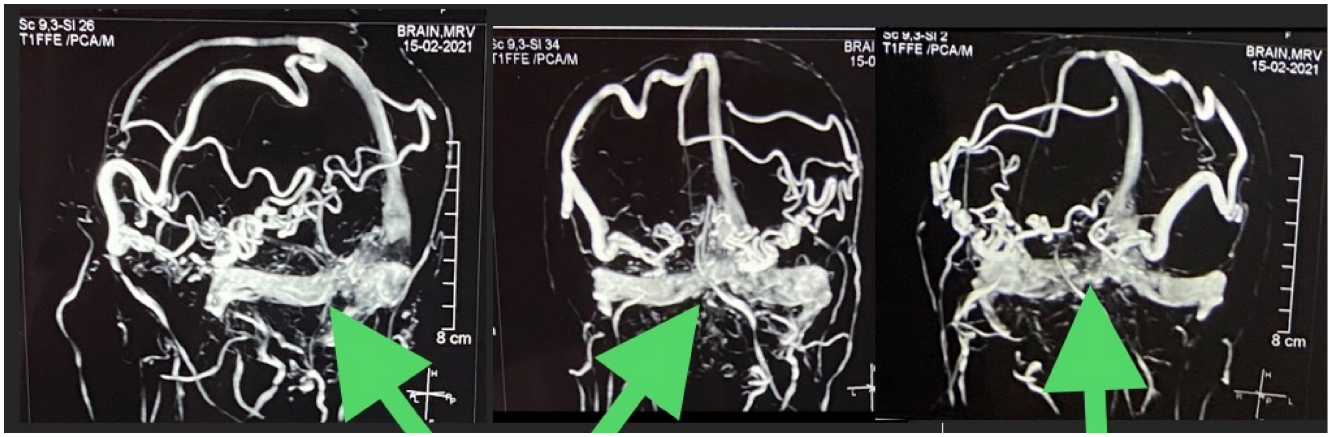

FIGURE 2 Lateral, anterior, and oblique views of MR venography showing heterogenous altered signal flow in bilateral transverse venous sinuses.

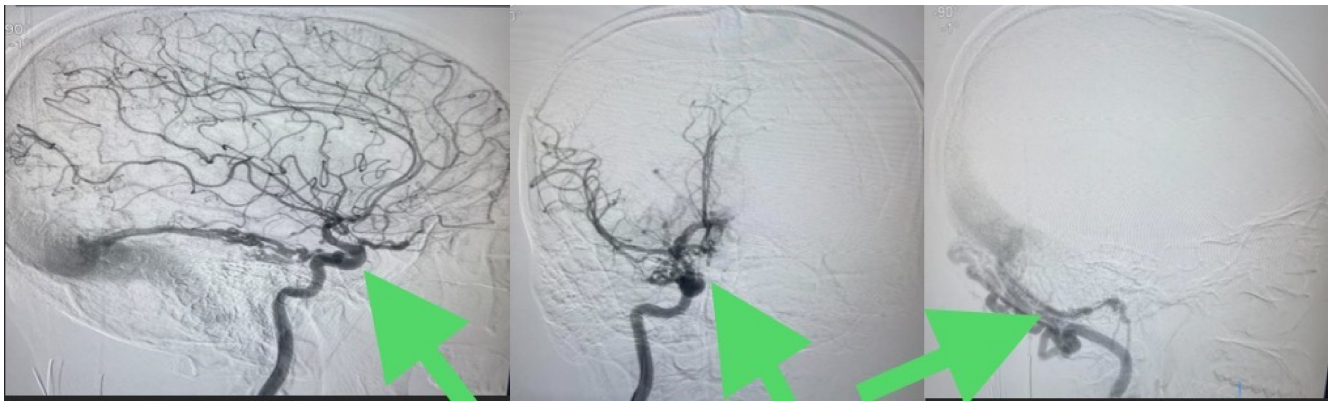

FIGURE 3 Lateral and anterior views of DSA showing bilateral giant arteriovenous fistula with multiple collaterals.

depends upon the extent, location, recanalization, and hypertension.[8] Most common clinical presentation of cerebral venous thrombosis is a headache (75%–95%).[9] Focal neurological deficits, seizures, altered mental status, and papilledema are the other common presenting complaints.[1] Focal neurological symptoms are more commonly seen in the patient with parenchymal changes as evidenced by imaging.[8] It has various clinical presentations varying from asymptomatic ones to serious clinical effects such as subarachnoid hemorrhage.[10]

It is difficult to diagnose only based on history and examination basis. Urgent neuroimaging is necessary to diagnose cerebral venous thrombosis. We preferred to use brain MRI and magnetic resonance (MR) venography or CT along with Cranial CT Venography if MRI is not available.[1] Most of the time CT head is non-specific. In some cases, CT demonstrates direct signs of CVT like dense triangle sign- hyperdense triangular shape area seen on the posterior part of superior sagittal sinus on non-contrast CT. Empty delta sign is a triangular shape also seen in the posterior part of sagittal sinus on contrast CT with lacking central contrast enhancement. Cord sign is also seen in contrast CT which is linear hyperdensity on the cerebral cortex.[11,12] CT head is generally normal in most cases so CT venography might be helpful to aid in the diagnosis in the absence of MR venography. CT venography help us by giving information about filling defects, sinus wall enhancement, and collateral venous drainage system.[8,13]

MRI with T2-weighted image in combination with MR venography is the most sensitive test to diagnose the CVT.[14] The MRI signal depends upon the age of the thrombus. Initial 5 days show isointense on T1 and hypointense on T2. After 5 days, it became apparent on both T1 and T2. After a month, it may show the variable pattern of the signal. We must know the different stages and their appearance on the different techniques to diagnose CVT. In the older thrombus, we may need intra-arterial angiography.[15]

Digital subtraction angiography (DSA) CT angiography is a last resort imaging technique to find out predisposing factors in CVT. Congenital anomalies like craniofacial venous abnormalities may predispose to the development of CVT. DSA CT angiography should be the preferred noninvasive modality to evaluate the case of CVT.[16] In our case, DSA was suggestive of bilateral dural arteriovenous fistula with multiple collaterals. Besides neuroimaging, American Heart Association recommends obtaining complete

blood count (CBC), chemistry panel, prothrombin time, and activated partial thromboplastin time for the suspected patient.[1] Investigation may reveal a hypercoagulable state, infection, and inflammatory process. Elevated D-dimer level supports the diagnosis of CVT but normal D-dimer levels do not rule out the diagnosis. Meta-analysis of 14 studies with a total of 1134 patients shows D-dimer was good with a sensitivity of 93.9% (95% CI 87.5–97.1) and specificity of 89.7% (95% CI 86.5–92.2).[17] Generally protein C, Protein S along anti-thrombin screening should be done in patients with suspected CVT.

There is a frequent association between DAVF and CVT. A significant number of patients diagnosed with DAVF, ranging from 39% to 78%, also experience concomitant CVT. Lindgren et al.[18] in their study indicated that DAVF occurs in a minimum of 2% of patients diagnosed with CVT. DAVF can be found in various locations within the dura, but they are most frequently observed in the transverse and cavernous sinuses. These DAVFs are commonly associated with chronic CVT onset, older age, and male gender. In most cases, DAVFs related to CVT are identified either simultaneously with the CVT diagnosis or subsequently during follow-up assessments. In our case, it was an adolescent male who was found to develop DAVF during follow-up. Schuchardt et al.[19] in the study also highlighted the importance of screening CVT patients for DAVF using dynamic magnetic resonance venography (MRV). However, it is still unclear whether CVT is the cause or the result of DAVF. According to most researchers, venous thrombosis and venous hypertension are considered the primary factors leading to the development of arteriovenous fistulas. Venous thrombosis increases venous pressure, which then triggers the opening and growth of the venous microcirculation shunt system, ultimately forming abnormal arteriovenous connections known as fistulas.[5,20] However, there are differing opinions suggesting that CVT is more likely a secondary event following DAVF. These studies have found that most cases of CVT were located downstream or around the fistula, indicating that CVT may occur as a delayed event after the development of DAVF. It is proposed that CVT could be caused by turbulent blood flow resulting from the shunts, while venous hypertension leads to engorgement of cortical veins and stagnation of blood flow.[21,22] In our report, the determination of whether the DAVF was primary or secondary cannot be definitively made without baseline cerebral angiography.

Treatment of CVT should be started as soon as possible once it is confirmed. It consists of reversal of known factors if present, control of intracranial hypertension and seizures along with anticoagulation therapy. With the aim of recanalizing veins along with propagation of thrombus to the nearby veins, others body structure and preventing recurrence of CVT, anticoagulation therapy generally started with either low molecular weight heparin (LMWH) subcutaneously or unfractionated heparin intravenously.[23,24] Along with the benefits, there is a risk of anticoagulation therapy in the patient with CVT. There is an increased risk of hemorrhage in previously infarcted areas.[23]

The preferred treatment for DAVF with associated CVT is anticoagulant therapy. However, the approach to endovascular treatment of the fistula may vary depending on the individual patient's circumstances. Surgical intervention is considered a reasonable option when the DAVF has caused significant clinical symptoms or when it has resulted in CVT. For patients with DAVF who have minor symptoms (except those caused by trauma) or symptoms that have spontaneously improved, immediate surgical intervention is not recommended. In such cases, the fistula may close off without the need for surgical intervention. The decision regarding the choice of treatment—whether anticoagulation, endovascular treatment, or surgery—is based on careful evaluation of the patient's clinical presentation, the severity of symptoms, and the specific characteristics of the DAVF. The treatment approach should be individualized and take into account the potential risks and benefits for each patient.[25] Our case was managed with anticoagulant therapy only.

DAVF has the potential to develop as a lasting complication of CVT. It is possible for DAVF to remain without symptoms, leading to a lower estimated prevalence if ongoing imaging is not conducted. Following an episode of CVT, it is crucial to carefully assess whether DAVF is present by means of follow-up imaging. It is necessary to regularly monitor patients to determine whether the DAVF occurred before the cerebrovascular thrombosis or whether it was the other way around.

4 | CONCLUSION

This case report highlights the importance of considering CVT as a differential diagnosis in young adolescents presenting with long-term headaches. It emphasizes the need for early identification of risk factors associated with CVT in order to prevent long-term complications. Timely investigation and clinical suspicion are crucial in patients presenting with chronic symptoms such as vomiting, headache, and visual disturbances. Additionally, the coexistence of CVT and DAVF should be taken into consideration, particularly in patients with a history of CVT.

AUTHOR CONTRIBUTIONS

Man Bahadur Paudyal: Conceptualization; writing – original draft; writing – review and editing. **Madhur Bhattarai:** Writing – original draft; writing – review and editing. **Neha Mehta:** Writing – original draft; writing

– review and editing. **Niraj Gautam:** Writing – review and editing. **Bikash Baral:** Writing – review and editing. **Niraj Kumar Sharma:** Writing – review and editing. **Rashika Basnet:** Writing – review and editing. **Bardan Ghimire:** Writing – review and editing.

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The authors have no conflict of interest to declare.

DATA AVAILABILITY STATEMENT
All the required information is available in the manuscript itself.

CONSENT
Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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REFERENCES
1. Dmytriw AA, Song JSA, Yu E, Poon CS. Cerebral venous thrombosis: state of the art diagnosis and management. *Neuroradiology*. 2018;60(7):669-685.
2. Coutinho JM, Ferro JM, Canhão P, et al. Cerebral venous and sinus thrombosis in women. *Stroke*. 2009;40(7):2356-2361.
3. Krayenbuehl H. Thrombosis of the cerebral veins and sinus. *Acta Neurochir Suppl (Wien)*. 1961;Suppl 7:248-254.
4. Liu KC, Bhatti MT, Chen JJ, et al. Presentation and progression of papilledema in cerebral venous sinus thrombosis. *Am J Ophthalmol*. 2020;213:1-8.
5. Yang S, You R, Wu W, Wei Z, Hong M, Peng Z. Dural arteriovenous fistula complicated with cerebral venous sinus thrombosis. *World Neurosurg*. 2020;134:348-352.
6. Gordon DL. The diagnosis and management of cerebral venous thrombosis. *Handbook of Cerebrovascular Diseases*. 2nd ed. Revised and Expanded CRC Press; 2004:605-635.
7. Gautam N, Gajurel BP, Shah S, et al. Clinical profile and short-term outcome of cerebral venous thrombosis in a Nepalese population: a retrospective study. *Nepal J Health Sci*. 2022;2(1):64-71.
8. Leach JL, Fortuna RB, Jones BV, Gaskill-Shipley MF. Imaging of cerebral venous thrombosis: current techniques, spectrum of findings, and diagnostic pitfalls. *Radiographics*. 2006;26(Suppl 1):S19-S41.
9. Mehta A, Danesh J, Kuruvilla D. Cerebral venous thrombosis headache. *Curr Pain Headache Rep*. 2019;23(7):1-6.
10. Benabu Y, Mark L, Daniel S, Glikstein R. Cerebral venous thrombosis presenting with subarachnoid hemorrhage. Case report and review. *Am J Emerg Med*. 2009;27(1):96-106.
11. Virapongse C, Cazenave C, Quisling R, Sarwar M, Hunter S. The empty delta sign: frequency and significance in 76 cases of dural sinus thrombosis. *Radiology*. 1987;162(3):779-785.
12. Lee EJY. Signs in imaging radiology the empty Delta sign 1. *Delta*. 2002;224:788-789.
13. Rodellac MH, Alexandre K, Antoine F, et al. Thrombosis and multidetector CT angiography: tips and tricks. *Radiographics*. 2006;26:5-18.
14. Boukobza M, Crassard I, Bousser MG, Chabriat H. MR imaging features of isolated cortical vein thrombosis: diagnosis and follow-up. *Am J Neuroradiol*. 2009;30(2):344-348.
15. Isensee C, Reul J, Thron A. Magnetic resonance imaging of thrombosed dural sinuses. *Stroke*. 1994;25(1):29-34.
16. Lu L, Zhang LJ, Poon CS, et al. Digital subtraction CT angiography for detection of intracranial aneurysms: comparison with three-dimensional digital subtraction angiography. *Radiology*. 2012;262(2):605-612.
17. Dentali F, Squizzato A, Marchesi C, Bonzini M, Ferro JM, Ageno W. D-dimer testing in the diagnosis of cerebral vein thrombosis: a systematic review and a meta-analysis of the literature. *J Thromb Haemost*. 2012;10(4):582-589.
18. Lindgren E, Rentzos A, Hiltunen S, et al. Dural arteriovenous fistulas in cerebral venous thrombosis. *Eur J Neurol*. 2022;29(3):761-770. doi:10.1111/ene.15192
19. Schuchardt FF, Demerath T, Elsheikh S, et al. Dural arteriovenous fistula formation secondary to cerebral venous thrombosis: longitudinal magnetic resonance imaging assessment using 4D-combo-MR-venography. *Thromb Haemost*. 2021;121(10):1345-1352. doi:10.1055/s-0041-1723991
20. Gupta A, Periakaruppan A. Intracranial dural arteriovenous fistulas: a review. *Indian J Radiol Imaging*. 2009;19(1):43-48. doi:10.4103/0971-3026.45344
21. Chen JG, Li ZX, Zhang DF, Wang JY, Hou LJ. Cerebral venous sinus thrombosis complicated with acute development of dural arteriovenous fistula: a case report. *J Clin Neurosci*. 2017;44:225-226.
22. Tsai LK, Jeng JS, Liu HM, Wang HJ, Yip PK. Intracranial dural arteriovenous fistulas with or without cerebral sinus thrombosis: analysis of 69 patients. *J Neurol Neurosurg Psychiatry*. 2004;75(11):1639-1641.
23. Bousser MG. Editorial – cerebral venous thrombosis: nothing, heparin, or local thrombolysis? *Stroke*. 1999;30(3):481-483.
24. Benamer HTS, Bone I. Cerebral venous thrombosis: anticoagulants or thrombolyic therapy? *J Neurol Neurosurg Psychiatry*. 2000;69(4):427-430.
25. Liang H, Xu C, Xu J. Cerebral venous sinus thrombosis and dural arteriovenous fistula associated with protein S deficiency: a case series study. *BMC Neurol*. 2022;22(1):164. doi:10.1186/s12883-022-02693-3

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