

A case of cerebral venous thrombosis presenting as acute reversible visual loss: a rarely reported association

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ABSTRACT

Cerebral venous sinus thrombosis (CVST) is not an uncommon cause of stroke but very often unrecognized at initial presentation due to lack of clinical suspicion and thus frequently left untreated. CVST is a potentially serious condition which manifests with diverse clinical manifestations, from isolated headache to focal neurological signs and even coma. CVST usually takes place either an inherited thrombophilia or any acquired hyperviscosity state and thus prompting anticoagulation was regimen as is the cornerstone of successful treatment. We describe a 47-year-old woman who presented with recurrent bouts of vomiting in the post-operative period and later developed cortical blindness and asymmetric limb weakness. Magnetic resonance imaging (MRI) showed hyperintensity involving bilateral parieto-occipital corticomedullary junction. MR venography showed signal void in the superior sagittal sinus. She was diagnosed as CVST and achieved complete recovery with anticoagulation therapy. Bilateral occipital infarction as a consequence of cerebral venous thrombosis is a rare cause of visual loss. Thrombosis in the superior sagittal sinus was related to her cortical blindness and weakness. This case illustrates that cerebral venous thrombosis should be considered in cases of occipital vascular lesions leading to acute painless loss of vision prompting anticoagulation therapy which can improve the outcome significantly. Dehydration could be considered as a risk factor for development of CVST in appropriate situations.

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KEY WORDS: Cerebral venous sinus thrombosis; occipital infarction; cortical blindness; dehydration, MR venography

doi : 10.5214/ans.0972.7531.200311

Introduction

Cerebral venous sinus thrombosis (CVST) is a potentially serious but treatable condition and the incidence varies between 1-3% of all strokes.¹ Risk factors of developing CVST can be either acquired conditions (eg, surgery, trauma, pregnancy, puerperium, antiphospholipid syndrome, cancer, exogenous hormones) or various inherited thrombophilia. CVST presents with spectrum of non-specific clinical profile from headache to coma. Lack of clinical suspicion often masquerades the diagnosis of CVST, which although carries a better prognosis than other ischemic strokes, bears a mortality of 5-10%.^{2,3} The pathogenesis behind the manifestations of CVST include raised intracranial tension due to impaired venous drainage and focal neurodeficit from venous infarct/ hemorrhage.⁴ Early diagnosis and prompt anticoagulation therapy is the key in management. Cortical blindness associated with CVST is very unusual and literature review has further revealed only a handful of case reports.⁵⁻⁷ Here, we describe a rare case of reversible cortical blindness due to bilateral occipital infarction resulting from CVST.

Case report

A 47-year-old non-diabetic, non-hypertensive woman presented with gradually worsening occipital headache for two days followed by acute onset bilateral painless loss of vision and limb weakness. She developed all the symptoms during postoperative period following a vaginal hysterectomy. She mentioned several bouts of vomiting prior to the event. The weakness initially started on the right side of the body and progressed to the left side. There was no associated fever, convulsion, altered mental status, dysphasia and sphincter dysfunction. She

denied any history of recent trauma. She had two children with no history of spontaneous abortion, use of oral contraceptive pills (OCP) or complicated puerperium.

The patient was drowsy and dehydrated. Blood pressure was 100/70 mm Hg, pulse rate 110/min, low volume and regular. On neurological examination, power in right upper and lower limbs was 2/5, in left lower limb it was 3/5 and in left upper limb was 4/5. Increased tone and brisk deep tendon reflexes were present in all four limbs. Plantar reflex was extensor bilaterally. On ophthalmological examination visual acuity was reduced to perception of light in both the eyes. Both eyes exhibited normal pupillary reflex with no restriction in the movement of extraocular muscles. On indirect ophthalmoscopy, mild to moderate right disc edema and a slightly swollen left disc were documented (Figure 1). Other neurological examinations were unremarkable. Her routine blood parameters were within normal limit. A non-contrast computed tomography (CT) scan of brain showed well defined hypodense areas in bilateral parieto-occipital border zones involving both gray and white matter not restricted to arterial territory. Adjacent cortical sulci are effaced suggestive of edema (Figure 2A). With the clinico-radiological suspicion of CVST D-dimer assay was done, which was 1705.17 ng/mL (N <500 ng/mL). Magnetic resonance imaging (MRI) of brain without contrast revealed areas of hyperintensity involving corticomedullary junction of bilateral parieto-occipital region (Figure 2B). Diffusion-weighted imaging (DWI) showed sharply delineated areas of diffusion restriction involving both gray and white matter of bilateral parietal and occipital

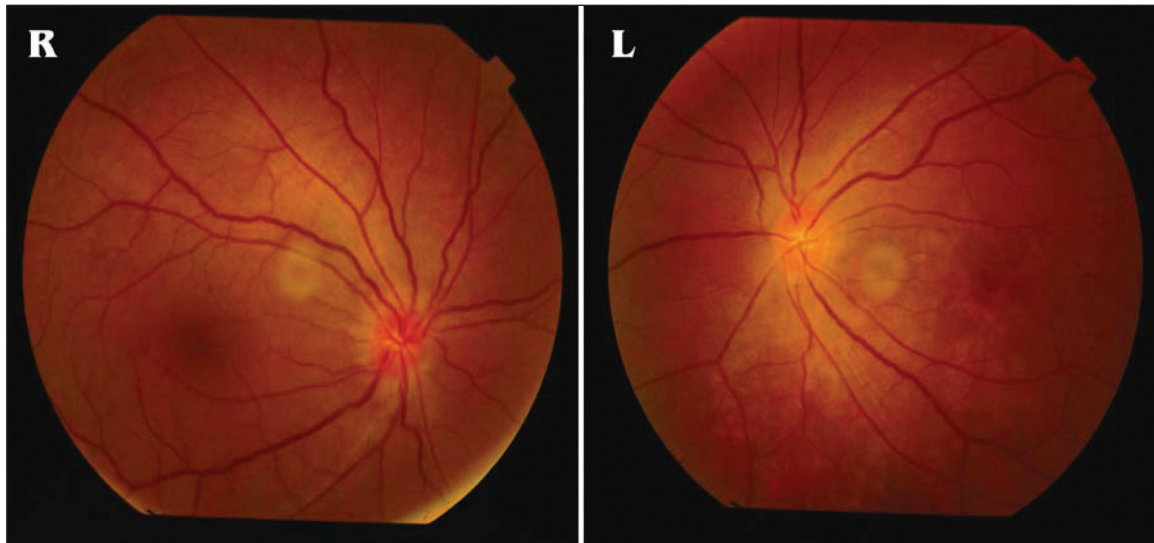


Fig. 1: Fundoscopic examination showing mild to moderate right disc edema and a slightly swollen left disc.

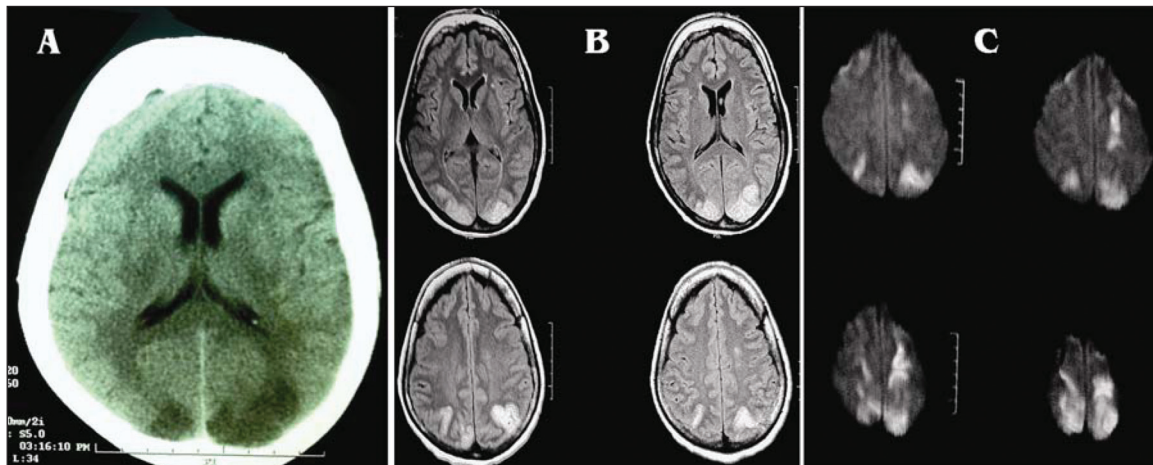


Fig. 2: (A) CT scan of brain showing well defined hypodense areas in bilateral parieto-occipital areas involving both gray and white matters. Adjacent cortical sulci are effaced suggestive of edema; (B) Axial T2-weighted FLAIR images showing areas of hyperintensity involving corticomedullary junction of bilateral parieto-occipital region; (C) Diffusion-weighted imaging (DWI) showed areas of diffusion restriction involving both gray and white matters of bilateral parietal and occipital regions.

regions (Figure 2C). Non-contrast MR venography revealed signal void in posterior aspect of superior sagittal sinus (Figure 3A). Non visualization of straight sinus, left transverse and sigmoid sinuses as well as left jugular bulb and internal jugular vein is also noted (Figure 3B). These findings, when corroborated with the spin echo, fluid attenuated inverse reconstruction (FLAIR) images, DWI and clinical profile of the patient is consistent with the diagnosis of cerebral venous sinus thrombosis with venous infarcts involving bilateral parieto-occipital areas in the border zone. An extensive search for existing hypercoagulable state was performed including protein C and S level, anti-nuclear antibody, anti-phospholipid antibody and homocysteine level but no abnormality could be demonstrated. Treatment with body weight adjusted low molecular weight heparin and oral anticoagulant therapy

resulted in marked recovery of vision and limb-weakness within one week and consequently the prothrombin time (PT) international standardized ratio (INR) reached the target range of 2 to 3. At 6-month follow up, she was symptom-free and follow-up MRI brain after six months revealed no residual abnormality. Study was conducted after obtaining an informed consent form from patient.

Discussion

(CVST) is a rare and potentially fatal type of cerebro-vascular disease which occurs in 5 individuals per million¹ with preponderance in female and younger age-group and if not treated properly, 4.3% of patients die during the acute phase of CVST.² There are many risk factors of CVST including hereditary thrombophilia (like factor V Leiden mutation, prothrombin-gene

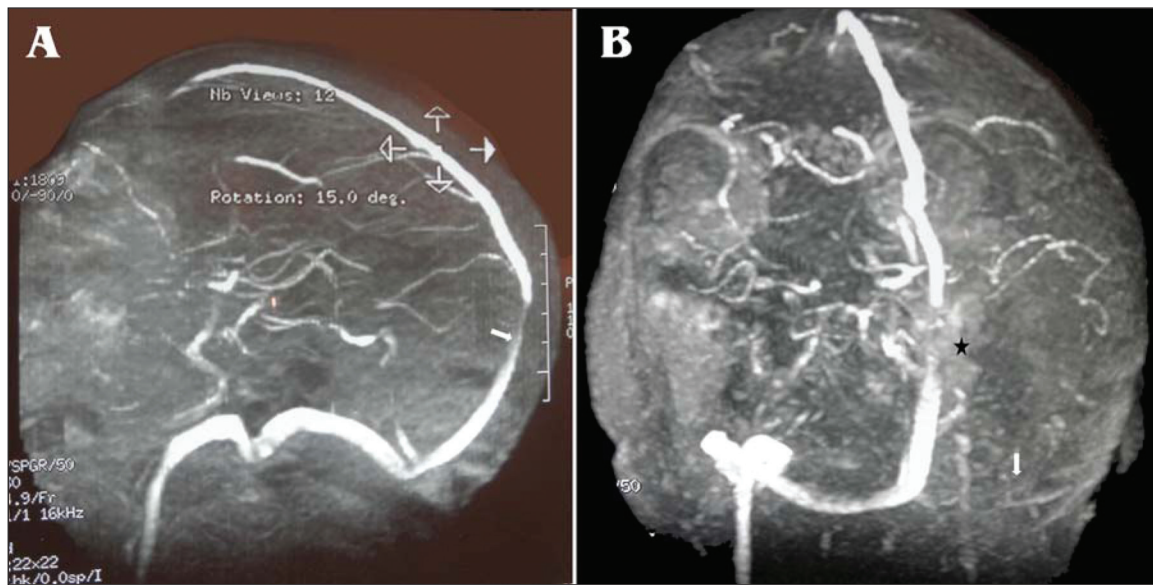


Fig. 3: (A) Non contrast MR venography on sagittal section revealed signal void (white arrow) in posterior aspect of superior sagittal sinus. (B) Coronal section showing signal void (black star) in superior sagittal sinus and non visualization of straight sinus, left transverse and sigmoid sinuses as well as left jugular bulb and internal jugular vein (white arrow) is noted.

mutation 20210GA, antithrombin III, protein C and S deficiency), various acquired hypercoagulable and hyperviscosity states (hyperhomocysteinaemia, antiphospholipid antibody syndrome, Behcet's disease, hematological disorders), pregnancy and puerperium, post-operative period, local or systemic inflammation or infectious processes, malignancies, drugs like oral contraceptives.⁸ It has been seen that 44% patients with CVST had one risk factor, 33% had two, 6% had 3 or more risk factors and in 17% patients no risk factor could be elucidated.³ Headache, diffuse and often severe, is the most common symptom of CVST. Seizure is found to be more frequent than in arterial stroke. Todd's paresis, if alternating between both sides of bodies, in adults, indicates a strong possibility of CVST, if not proved otherwise. CVST can present as any of the following, (1) central motor or sensory deficits, aphasia and other neuro-deficits (40-60%), (2) syndrome of isolated intracranial hypertension with headache, vomiting and blurred vision owing to papilloedema (20-40%) and (3) impaired consciousness (10-20%).⁹ Ophthalmological manifestations of CVST are rare and are primarily the consequence of increased intracranial pressure. If cerebral infarction involves the parieto-occipital (P-O) region, visual disorientation, psychic gaze paralysis and inferior altitudinal hemianopsia may develop.¹⁰ Unlike, deep vein thrombosis (DVT) of lower limbs, role of D-dimer in CVST is controversial because it is elevated in neurodeficient CVST but low in CVST with isolated headache.¹¹

Superior sagittal sinus alone or in combination with lateral sinus is the most common site of CVST. Computer Tomography (CT) scan of brain is the initial investigation. Although 'dense triangle sign', 'cord sign' or 'empty delta sign' in CT are classical of CVST, it usually presents with non specific changes like brain oedema and hemorrhagic infarcts involving border zone. (MRI) of brain is much more sensitive than CT in this condition and often demonstrates the clot, unless the scan is obtained during the first few days or months later. CT venography or MR venography may be used depending upon availability for the

diagnosis, staging and follow-up.^{12,13} Two- or three-dimensional time of flight (TOF) MR angiography or gadolinium-enhanced three-dimensional auto-triggered elliptic centric-ordered (AT-ECO) MR venography are among some modalities for imaging the anatomy of cerebral venous system with further precision.¹⁴

The mainstay of managing CVST is either activated prothrombin time (aPTT) adjusted intravenous heparin or body weight adjusted subcutaneous low molecular weight heparin (LMWH), which prevents propagation of thrombotic process and prevents re-occlusion of endogenously re-canalised vessels. Once the level of consciousness is normalized therapy is switched over to oral anticoagulants aiming to keep the INR between 2 to 3 and to continue for 3 months in idiopathic CVST, 3 to 6 months in perinatal period or with the use of oral contraceptive pills (OCP) and 6-12 months in case of hereditary thrombophilia.¹⁵ Prophylactic antiepileptics are recommended when there are associated focal neuro-deficits or focal parenchymal lesion in neuro-imaging and conventional modalities in order to reduce intracranial tension these are indicated in papilloedema and threatened vision. Role of endovascular thrombolysis and clot removal by balloon angioplasty are also being studied in randomized control trial. With prompt anticoagulation and long-term follow-up, independent survival is the rule albeit some degree of neuro-deficit is persistent in the surviving patients.

In the present case there was possible contribution of dehydration in the causation of CVST. Acute onset cortical blindness probably occurred as a consequence of bilateral occipital venous infarction. It is probably related to superior sagittal sinus thrombosis. The limb weakness in the patient was thought to be due to bilateral parietal cortex involvement from thrombosis of superficial cerebral veins. Thrombosis involving deep venous system may involve thalamus or basal ganglia which may also contribute to the limb weakness. Although areas of diffusion restriction, involving corticomedullary junction of bilateral parieto-

occipital border zone, in diffusion weighted images corroborates with presence of venous infarcts, minimal papilloedema may be contributed to some degree of vasogenic edema.

Conclusion

Our case is unique in the way that cerebral venous sinus thrombosis presented with cortical blindness probably due to bilateral occipital infarct and remarkable recovery of symptoms on prompt therapy with anticoagulants. It is evident from the discussion that, albeit, venous sinus thrombosis mimics other cerebro-vascular events clinically, may even radiologically, the philosophy of management is completely different. High index of suspicion along with newer radiological techniques is needed to clinch the diagnosis at the earliest and early anticoagulation can successfully overcome this dreadful yet reversible condition.

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