Cerebral Venous Sinus Thrombosis Presenting with Features of Raised Intracranial Tension and En Plaque Meningeal Thickening; A Case Report

Sir,

Chronic venous sinus thrombosis can present with different symptoms. Headache, transient visual obscuration, diplopia are common symptoms developing because of raised intracranial tension. Venous sinus thrombosis needs to be ruled out in every case with features suggestive of raised intracranial tension. Seizures, cranial nerve palsies, limb weakness, mental status changes can be seen when there is parenchymal involvement. Radiological finding of venous thrombosis includes visualization of thrombus, irregular sinus lumen manifesting partial obliteration by thrombus, parenchymal abnormalities in form of venous haemorrhagic infarct and edema. Subarachnoid and rarely subdural haemorrhage can also occur. Deep vein thrombosis and isolated cortical vein thrombosis are unusual findings of cerebral venous thrombosis. Parenchymal contrast enhancement is reported in up to 29% of cerebral venous thrombosis and this is typically gyral in location.[1] Meningeal thickening and enhancement is a very rare manifestation of cerebral venous thrombosis.[2] We report here a case of cerebral venous thrombosis presenting with features of raised intracranial tension and localized pachymeningeal thickening and enhancement.

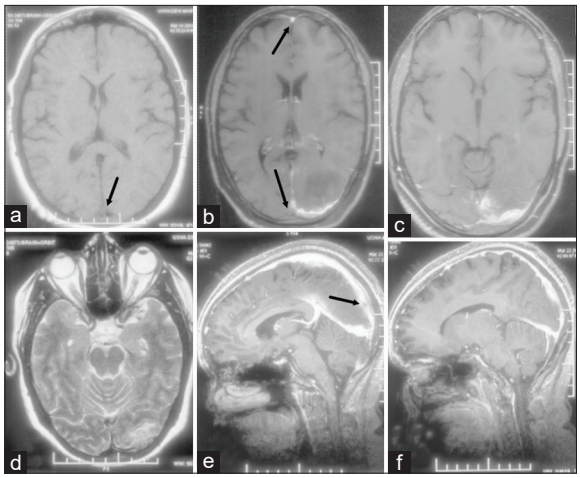

Figure 1: (a) T1 axial sequence showing partial absence of flow voids in posterior sagittal sinus- Arrow. (b.) Post contrast T1 axial sequence showing pachymeningeal thickening and post contrast enhancement in left occipto-parietal convexity. Hypointensity is seen in left occipital lobe white matter s/o edema. Contrast is seen filling anterior part of sagittal sinus but not the posterior part- Arrows. (c) T2 axial sequence showing white matter hyperintensity s/o edema in left occipital lobe and partial absence of flow voids in posterior sagittal sinus. (d) Post contrast T1 sagittal sequence showing pachymeningeal thickening and post contrast enhancement. Partial filling defect is seen in posterior part of sagittal sinus- Arrow

A 44-year-old woman was admitted in April 2018 with seven months' history of episodic hemi-cranial headaches and three months' history of mild visual diminution. Headache was episodic, occurring every 2-3 days and was lasting 4-6 hours. It was predominantly left hemi-cranial and occasionally occurred on right. It was moderate in intensity with visual analogue scale of 5. The headache was associated with occasional nausea but there was no vomiting, photophobia or phonophobia, watering/congestion of eyes or nose. There was no fever, neck pain or alteration of consciousness. No h/o ear discharge, earache or tinnitus was there. There was associated mild diminution of vision in both eyes for last 3 months. This was insidious in onset and was noted as blurring of far placed objects. Blurring has been improving for last 1 month but it had not improved completely. Transient visual obscurations were denied. No desaturation of colours, pain in eye while movement or prominence of eyeballs was there. No double vision or deviation of eye was there. There was no history suggestive of any other cranial nerve involvement apart from visual diminution. There was no higher mental function, extrapyramidal, motor, sensory or autonomic involvement. There was no h/o weight gain/loss, night sweats, anorexia, menstrual irregularity. There were also no h/o joint pains, oral/genital ulcers, dry eyes/mouth. There was no history suggestive of any other systemic illness. Her menstrual history was normal and she was not on any treatment apart from intermittent pain killers. She has had history of similar headaches few years back and she had transient double vision also, this

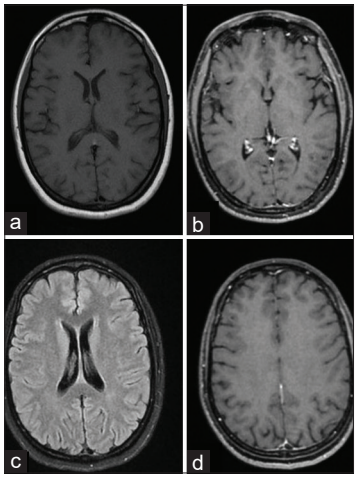

Figure 2: (a) T1 axial sequence showing normal flow voids in venous sinuses. (c) FLAIR axial sequence showing normal parenchyma and normal flow voids in venous sinuses. (b and d) Post contrast T1 axial sequence showing mild post contrast enhancement in pachymeninges along posterior aspect. Abnormal thickening is not seen

was resolved in few days. No documentation of treatment was available, though MRI brain was available and it was reviewed and found to be normal. On examination at admission, she was fairly built and nourished, general examination was unremarkable. Visual acuity was 6/9 in both eyes. There was no relative afferent pupillary defect. Papilledema was present in both eyes. No extraocular movement restriction was there. Rest neurological examination was normal. There were no meningeal signs. Her MRI was available, done two weeks prior to presentation and it is presented in Figure 1. It was showing superior sagittal sinus and left transverse sinus thrombosis. There was also pachymeningeal thickening with post contrast enhancement in left occipito-parietal convexity and posterior falx. There was also feature suggestive of raised intracranial tension in form of partial empty sella. The meningeal thickening was deemed as en plaque meningioma. Her investigations including hemogram, renal function, liver function, sugars, thyroid profile, viral markers, ANA/dsDNA, ANCA, anticardiolipin were unremarkable except for Hb of 10.1 gm/dl and low vitamin D levels. Chest radiograph PA view was unremarkable. Given the high possibility of meningioma and secondary venous sinus thrombosis, neurosurgery intervention was sought and no further tests were done at this point of time. She was started on anticoagulation treatment (vitamin K antagonist- Acitrom 3 mg OD) and advised to keep international normalized ratio between 2 to 3. She was also advised about dietary restrictions which needs to be implemented along with vitamin K antagonist therapy. Acetazolamide and supplements like iron, calcium and vitamin D were started. She was discharged with advice to follow up with neurosurgery for biopsy and/or excision of the meningeal lesion. She was continued on anticoagulation and symptomatic treatment and her headache was improved. Though, there was further deterioration followed by stabilisation of visual acuity at 6/24 in right eye and 6/18 in left eye. She was admitted under

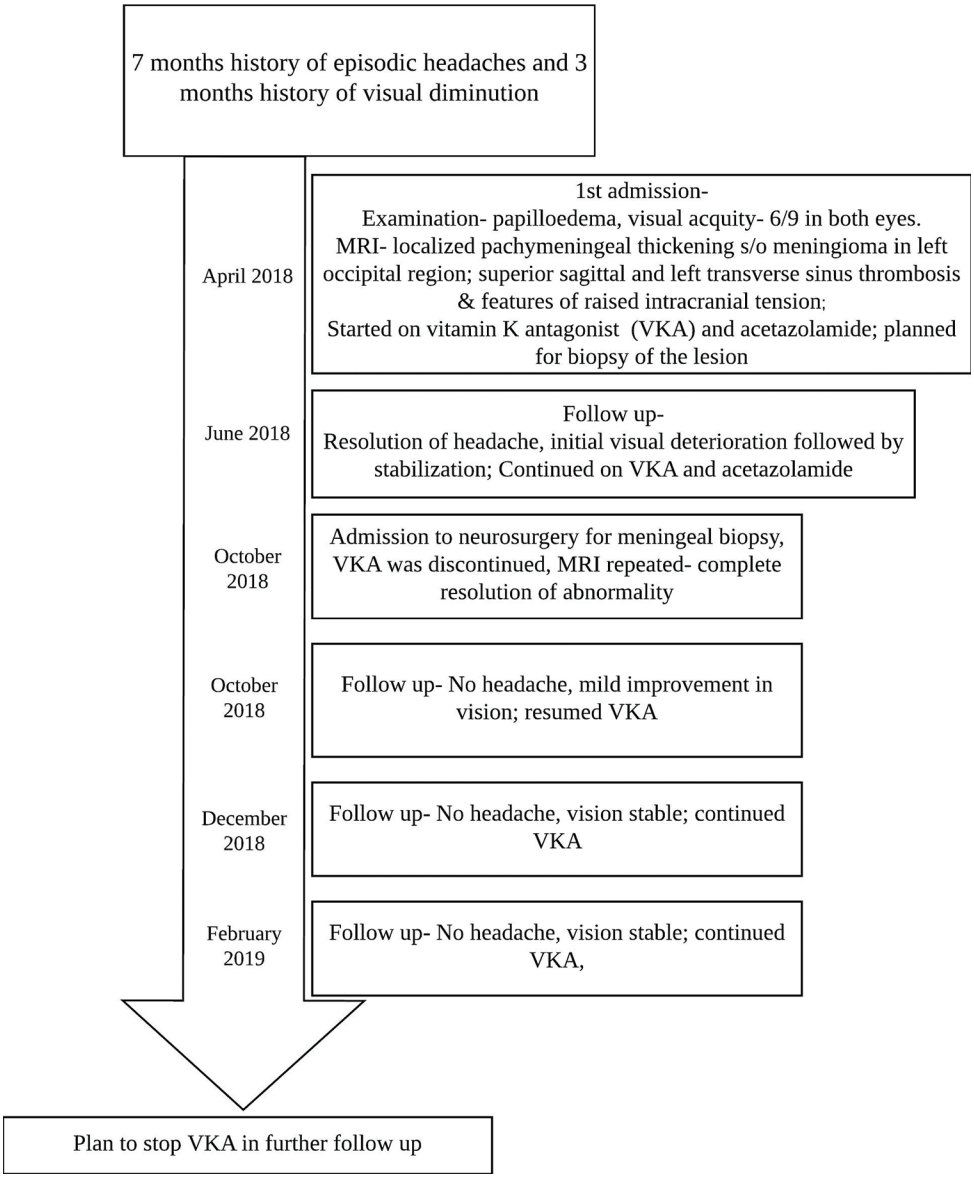

Figure 3: Chronology of events

neurosurgery department in October 2018 and MRI was repeated. Figure 2. It showed complete resolution of meningeal thickening, there was just mild post contrast enhancement. There was narrowing of left transverse sinus but sinuses had recanalized. Anticoagulation was discontinued as surgery was planned but it was resumed as there was no lesion. She was last followed up in February 2019, she did not have headache and no further deterioratin in vision was there. Plan is to stop anticoagulation in follow up and to send pending work up for thrombophilia including protein C, S and antithrombin. Figure 3 demonstrates the chronology of events.

Pachymeningeal enhancement without thickening is seen in half of the patients who undergo post contrast studies. This enhancement is of less than 2 mm thickness and seen normally in regions of dural reflections e.g., falx, tentorium and along anterior portions of anterior temporal lobe.[3,4] The vessels in dura mater do not produce a blood brain barrier so that the contrast material can leak out of blood vessels.[5] Pathological enhancement is characterized by increased meningeal intensity and thickening with or without nodularity. Intracranial hypotension, infections, autoimmune conditions such as sarcoidosis, vasculitides such as wegner's granulomatosis, iatrogenic such as post lumbar puncture and neoplastic diseases are common causes of pathological pachymeningeal enhancement. When thickening is localised, neoplastic process e.g., meningioma or inflammatory process is the most likely possibility. When no cause is apparent after thorough investigations, it is labelled as idiopathic hypertrophic meningitis.[6] Cerebral venous thrombosis as a cause of pachymeningeal thickening and enhancement is reported scantily.[2,7] On the contrary, pachymeningitis as a cause of venous sinus thrombosis is reported more commonly.[8,9] In the given case, it might be unclear whether pachymeningeal thickening is the cause or the consequence of the venous sinus thrombosis. Clinical presentation and regular follow up might give clue on this. In our case, though meningioma and secondary venous sinus thrombosis was the first possibility initially and we had planned for meningeal biopsy, we could have done other investigations to look for other causes of pachymeningeal thickening. CSF analysis with cytology for malignant cells, work up for sarcoidosis and behcet's disease could have been done at the time of admission. Patient was followed regularly and there was no further deterioration in her condition and in fact the headache was gradually diminished. On repeat MRI done prior to planned surgery, complete resolution of lesions leaded us to think that venous sinus thrombosis was the cause and not the consequence of pachymeningeal thickening as we did not treat anything apart from giving anticoagulation and symptomatic treatment.

Our patient highlights the cerebral venous thrombosis as a cause of pachymeningeal thickening and enhancement. Nevertheless, diligent look into other causes of pachyemeningitis and close follow up with periodic examinations are of utmost importance for establishing correct diagnosis. This may also avoid invasive procedures thus ultimately benefiting the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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