CEREBRAL VEIN AND DURAL SINUS THROMBOSIS

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ABSTRACT

We present a case of 29 years old female patient with 20 days history of headache, projectile vomiting and low grade fever. She was referred from Swat where she was locally treated as acute pyogenic meningitis with no improvement. Patient had right 6th cranial nerve palsy and papilledema. CT brain was unremarkable and MRI brain with MR Venography showed superior sagital, confluence of sinuses and transverse sinus thrombosis.

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INTRODUCTION

Cerebral vein and dural sinus thrombosis (CVST) is a rare disorder with a very low incidence¹. The estimated prevalence is 3 to 4 cases per one million population and up to 7 cases per 1 million population in children². While in a recent study in Iran, the estimated frequency was reported to be a bit higher with an incidence of 12.3 per 1 million population³. Cerebral vein thrombosis (CVT) is more common in young patient particularly in women and the mean age at diagnosis is 39 years⁴. Exact pathophysiology of CVT is unknown. Two mechanisms have been proposed. CVT resulting in local effects of vasogenic edema, parenchyma lesion like infarct or hemorrhagic infarction. Thrombosis of dural sinus results in impairment of CSF absorption and raised intracranial pressure⁵.

Clinical features of CVST are highly variable and do not help in diagnosis⁶. CVST symptoms can be assorted into several groups (syndrome). Important syndromes include isolated intracranial hypertension (headache, vomiting, papilledema, visual disturbances, sixth cranial nerve palsy) focal neurodeficits (monoparesis, hemiparesis, sensory impairment and seizures), encephalopathy with multifocal signs, altered level of consciousness and diffuse pattern of neurological deficits and cavernous sinus thrombosis (with ocular signs like chemosis, proptosis, occultor moter nerve palsy, facial pain)⁷. The most common risk factors are acquired and genetic prothrombotic conditions including protein C, protein S & anti thrombin III deficiency, Nephrotic Syndrome, anti phospholipid antibody syndrome, Factor V Laden mutations, pregnancy and puerperium, oral contraceptives pills and infections⁸,⁹. However 37% of patient don’t have any identified risk factors¹⁰.

The initial study usually performed in diagnosis of CVST is CT brain, the classic signs on CT brain is empty triangle sign, delta sign and cord sign¹². However CT can be normal in 30% of patients. MR imaging along with MR Venogram is the most sensitive diagnostic tool in diagnosis of CVST¹³. Aside from neuroimaging there is no lab test that can confidently rule out CVST. An elevated D-Dimer support the diagnosis but a normal value does not exclude it¹⁴. Examination of CSF should also be included in evaluating case of CSVT in addition to Lab test for prothrombotic conditions.

Once CVST has been diagnosed, immediate treatment should be instituted. Goal of treatment is to prevent propagation, treat underlying thrombophilia, and to recanalize the cerebral vein and dural sinus. These objectives can be achieved with either LMW Heparin or unfractionated Heparin¹⁵. Oral Anticoagulant should be started with target INR of 2-3. The duration of oral therapy depends upon underlying condition. Three months treatment is required when underlying prothrombotic condition is transient and corrected. In idiopathic condition treatment duration is 6-12 months, while in patients with severe thrombophilia life long treatment with oral anticoagulant is required.

CASE REPORT

A 29 years old female patient from Swat married with two issues present to us with 20 days history of headache, which was unilateral initially involving left side of head and later become generalized, vomiting which was projectile and usually occur early in the morning without any association with food intake. She was also complaining of Fever low grade, continuous in nature. Patient was not having any significant medical or surgical history. Her socioeconomic status was satisfactory and she was only using oral contraceptive pills.

Patient was seen at the start of these symptoms by local physicians in Swat where she was diagnosed
as acute pyogenic meningitis after Lumber Puncture was performed. She was started on intravenous ceftriaxone but the patient continues to have above symptoms. She was referred to Peshawar.

Here on arrival, patient was giving history of above symptoms. On examination patient was having right sided sixth cranial nerve palsy. She was having papilledema on fundoscopy. CT brain was performed that was non informative. MRI brain was then performed along with MR Venography that showed superior sagital, confluence of sinuses and transverse sinus thrombosis (Figures 1-3). Patient was diagnoses as a case of dural sinus thrombosis and was started on LMW Heparin and warfarin.

**DISCUSSION**

In this case report, patient present with signs and symptoms of raised intracranial pressure. Patient was having headache, Projectile vomiting. Fundoscopic examination was associated with Papiledema. Patient was having right sided 6th cranial nerve paralysis. Symptoms of cerebral vein thrombosis can be grouped into four categories and one among them is presentation with raised intra cranial pressure, resulting in headache vomiting, papilledema and cranial nerve sixth palsy. Headache can sometime be the only symptom and precede other symptoms by days or weeks. The other groups of symptoms with which the patient can present is Focal symptoms, Encephalopathy and cavernous sinus thrombosis.

CVST is more common in women, may be because of the effects of pregnancy and puerperium as well as because of use of OCPs. As compare to arterial type of stroke it is more common in young patient, mean age at diagnosis is 39 years, which is much younger then the age group for arterial type of stroke. There are several risk factors for CVST, in addition to procoagulant state which can be genetic or acquired, Pregnancy, OCPs, Infections (meningitis, otitis, mastoiditis, sinusitis, neck face mouth infection, systemic infections) head trauma, hematological disorder like PRV, PNH etc. Malignancy (CNS, systemic malignancies) can be some of the risk factors.

Patient presented to us had several risk factors; she was using oral contraception’s, as well as she was initially diagnosed as having acute pyogenic infection upon lumber puncture. However it is worth mentioning that a significant number of patients may present with CVST without having any identifiable risk factor. While the most frequent identified risk factors are prothrombotic state either genetic or acquired, pregnancy and puerperium and infections. Differential diagnosis, include but not limited to stroke due to arterial pathology, meningitis, encephalitis, brain abscess, cerebellar vasculitis.

CVST should be differentiated from arterial type of stroke, as the latter usually occur in old age, have a sudden onset, and not usually associated with seizures, and
raised intracranial pressure. Even in patients with CVST who has cerebellar infarct, territory of infarct does not follow arterial distribution. Diagnosis of CVST can be achieved with accuracy with MRI in combination with MR Venography. However CT is the investigation most commonly performed initially in clinical settings to rule out other cerebral acute or subacute disorders. CT will be abnormal only in about 30% of patients, showing dense triangle sign, empty triangle sign and cord sign. A great majority of patients will have normal CT Brain reported as was the patient admitted with us.

Patient should be evaluated for other prothrombotic states via protein C, protein S levels, Factor V leiden mutation, Anti-phospholipid antibody syndrome, hyperhomocystonemia, which was all within normal limit in patients admitted to us. Patient should be discouraged using combine contraceptive pills, instead other methods of contraception should be considered. An important point in CVST is the subsequent pregnancy, since CVST per say include pregnancy in its risk factors. However in patient without any thrombophilic states, pregnancy should not be considered as a contraindication to pregnancy. Our institution recommends treating pregnant patient with history of CVT with either low molecular heparin or unfractionated heparin during 3rd trimester, till eight week of delivery. Women should be advised not to get pregnant while on Warfain.

REFERENCES