AmbitJournals

Ambit Journal of Neuroscience and Behavioral Health. Vol 5(3) pp. 146-150. September 2019.
https://doi.org/10.32619/ajnbh

Full Length Research Paper

Cerebral venous sinus thrombosis is presenting as unusual bilateral blindness: A rare association case report

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Accepted 8th August 2019.

Cerebral venous sinus thrombosis (CVST) is an uncommon yet perilous condition. CVST symptoms are mainly severe headaches but other complications may occur, like weakness on one side of the body, difficulty in speaking or seizures and changes in mental status, however, we are reporting a cerebral venous sinus thrombosis case of a 44 years old Caucasian female, that was admitted to conservative therapy for an onset of severe, ongoing headache and blindness that affected both eyes. The patient was improved upon hospitalization and prompt anticoagulation therapy. Despite rare vision impairment was reported, our case report shows an unusual association of bilateral blindness and CVST that was not described before in the medical literature.

Keywords: cerebral venous sinus thrombosis, superior sagittal sinus, bilateral blindness, computed tomography.

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INTRODUCTION

Cerebral venous sinus thrombosis (CVST) is a distinct cerebrovascular disorder. The symptoms and complications occurring are highly variable. Women are more prone to CVST than men. About two-thirds of adult patients are women. Mostly women who have had recent headaches after starting oral contraception or women who have had seizures after giving birth in the obstetrical ward may have sinus thrombosis [1]. The clinical presentation of CVST is varied and may include headache, vomiting, seizures, unexplained changes in status, and a depressed level of consciousness [2]. In this article, we report a case of a patient who developed unusual bilateral blindness accompanied to cerebral venous sinus thrombosis. It is hoped that the presentation of this case will draw attention to this unexpected rare and devastating symptom and that this condition will put into consideration. Hence, educating clinicians and increasing their awareness of the unusual symptom that resulted due to CVST.

Case presentation

A 44yrs old female patient, with no chronic medical disease, was presented to the hospital with chief complaints of 1 Month history of headache (in the right side of the head), otitis media. The patient described the headache as, “the worst headache of her life.” The patient stated the headache was in the occipital and frontal areas and characterized the pain as a dull pain. The symptoms are associated with tinnitus, neck spasm, photopia, confusion. No nausea or vomiting was associated with the case.

Early morning the day she got presented to the hospital, the patient started to experience bilateral loss of vision and confusion. There was no history of nausea, vomiting, seizure, fever or bleeding from anywhere. There is no family history of blood disorder. The patient has primary infertility but hasn’t received any hormonal therapy in the last 2 years. The patient received hormonal therapy for her primary infertility. The patient is a smoker. she has a history of substance abuse. Surgical history was 4 tubal fertility surgeries (last surgery was 5 years ago). Physical examination revealed normal vital signs and the Neurological examination showed confusion, bilateral loss of vision (except projection of light), kernig’s sign and brudzinski’s sign , extra-ocular movement EOM were negative , pupils symmetrical equal reactive .

However, papilledema was noted on the physical exam.

Over the course of hospitalization was placed on IV fluids, oxygen, and tramadol for pain, IV heparin dose that was adjusted according to the patient’s body weight. The coagulation profile daily measured Then warfarin was added until INR was targeted 2-3. Omeprazole and paracetamol were also prescribed. After 2 days from treatment, the patient showed significant improvement from 8/10 severity to 4/10 from headache, conscious, alert, orted .

Blindness was improved and she was able to read with visual acuity (VA) 20/20.

Lab results stated that D- dimer was marginally raised. Toxic screen and test, thrombin time and lupus anticoagulant, anti-cardiolipin antibody, anti-β2 glycoprotein 1 antibody, activated protein C resistance were negative.

CT brain showed delta sign picture of cerebral sinus thrombosis. CTV showed thrombosis of superior sagittal sinus thrombosis (SSST), right transverse and right sigmoid sinuses. CTA and MRI were normal, MR Venogram Brain and MR angiography Head stated that no acute infarction was found. However, left cerebellar hemisphere tiny foci of subacute infarction were noted. MR venogram and MR angiography stated that there was no acute infarction. However, a tiny focus of subacute infarction in left cerebellar hemisphere was found. MRV shows no visualization of venous flow in superior sagittal sinus which may indicate venous sinus thrombosis and an attenuated right transverse sinus flow.

DISCUSSION

(CVST) is a rare and potentially serious type of cerebrovascular disease that is more predominant in females. It is estimated that CVST occurs in five individuals for every million people [3]. If not treated in the correct way, mortality rate would be 4.3% of CVST patients in the acute phase [4]. Despite that hereditary thrombophilia, intracranial, local infections, pregnancy, puerperium, postoperative state, and the use of oral contraceptives or hormonal therapy are considered as the main risk factors of CVST, less than 20% of the cases are of no known clear risk factors [5]. One of the most common symptoms of CVST is severe headache. In many cases the headache is the only symptom of CVST. Each 9 out of 10 individuals with sinus thrombosis have a headache; that may develop suddenly or tends to worsen over a period of several days [6]. Other symptoms may include seizures that are more common in women who develop sinus thrombosis, postpartum, difficulty to move one or more limbs, weakness on one side of the body, speech difficulties, mental status alteration and depressed level of consciousness. Superior sagittal sinus alone or in combination with lateral sinus is the most common site of CVST. Ophthalmological manifestations of CVST are not common and are mainly the consequence of elevated intracranial pressure (ICP). Papilledema is manifested sometimes asymmetrically in the 2 eyes and rarely in 1 eye only [7]. Computed Tomography scan of brain is the initial tool for investigation. Delta sign in CT is classical of CVST, it usually presents with nonspecific changes like brain oedema or elevated intracranial pressure. (MRI) of brain is much more sensitive than CT in this condition and often demonstrates the thrombus, unless the scan is obtained during the early few days or months afterward. CT venography or MR venography could if available for the diagnosis, staging, and follow-up [8]. MR angiography or MR venography are among some modalities for imaging the anatomy of cerebral venous system with further precision [9]. The main pillar of

managing CVST in patients with no contraindications for anticoagulation (AC) is either by body weight-adjusted subcutaneous low molecular weight heparin (LMWH) or activated prothrombin time (aPTT) adjusted intravenous heparin [10]. The rationale of anticoagulant therapy in CVST is to avoid thrombus extension and to favour spontaneous thrombus resolution. When the level of awareness is adjusted to normal, therapy is switched to oral anticoagulants aiming to keep the INR between 2 to 3. Oral AC is prescribed for 3 months if CVST was secondary to a transient risk factor or 6–12 months in patients with idiopathic CVST [11]. Conventional modalities in order to decrease intracranial pressure are indicated in papilloedema and threatened vision. Acetazolamide might be considered if the papilloedema is persistent. In few patients, vision continues to deteriorate despite taking acetazolamide. In these cases, shunting procedures (lumboperitoneal, ventriculoperitoneal shunts or optic nerve fenestration) should be considered. With immediate anticoagulation therapy and long-term follow-up, survival rate increases even if some degree of neuro-deficit remains in the surviving patients.

In the present case, there was acute blindness in the causation of CVST. Unusual bilateral blindness occurred because of papilloedema that occurred upon hospitalization. It is probably related to superior sagittal sinus thrombosis (SSS) that caused optic disc congested and elevated Intracranial pressure. Severe headaches were treated with tramadol to alleviate pain, but it was not fully eliminated. The patient had tubal fertility 4 times and had hormonal therapy which might be the reason behind the development of the CVST. The limb weakness in the patient may be contributed to Cerebral venous sinus thrombosis, affecting superior sagittal sinus. The ongoing pain and persistent blindness need further investigations to manage the unexpected complication of CVST.

CONCLUSION

Our case is unique in the way that cerebral venous sinus thrombosis is presented with unusual bilateral blindness that was probably due to superior sagittal sinus thrombosis with improvement of symptoms on prompt therapy with anticoagulants. It is evident from the discussion that, although symptoms of CVST are known, an unexpected unique new symptom might appear that needs to be considered. Further investigations and research are still required in the pathophysiology and management of CVST to ensure the finest possible treatment for patients.

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