

Case Report:

Cerebral Venous Thrombosis Late Presentation: Critical Care Case Report

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Abstract:

Cerebral venous thrombosis is an important cause of stroke in the young. The most common symptoms are headache, seizures and focal neurological deficits. A case of 13-year-old Saudi boy developed Cerebral venous thrombosis (CVT)(1). Final diagnosis is partial occlusion of left transverse and sigmoid sinus in consideration of cerebral venous thrombosis, beside right mastoid inflammation process and sinusopathy. Further investigation revealed polymorphism of the hypofibrinolytic plasminogen activator inhibitor type 1 gene(1,2).

Introduction:

Cerebral venous thrombosis (CVT) is less frequent than ischemic stroke or intracerebral hemorrhage. Cerebral venous thrombosis is an important cause of stroke in the young(3). The most common symptoms are headache, seizures and focal neurological deficits. The diagnosis can be confirmed with MRI, CT-venography and less commonly use catheter angiography. Approximately 40% of patients with CVT also present with intracranial hemorrhage(4). The use of oral contraceptives, and pregnancy are risk factors for cerebral-vein thrombosis, no information about the relation between the disorder and the prothrombin-gene mutation is yet available, recently a significant association was demonstrated between CVT and all inherited thrombophilia factors, as well as increased levels of homocysteine(5).

Case Report

13-year-old Saudi boy presented to tertiary hospital emergency department having experienced worsening headache with confusion and decreased vision for 7 days. Associated with intermittent fever, nasal obstruction and vomiting for 5 times. 1 week before admission, a left eye convergent strabismus became prominent, leading the patient's parents to emergency department. Physical examination revealed left abducens nerve palsy, with secondary diplopia within the extreme direction of gaze, bilateral papilledema and decreased visual acuity. (CT) contrast of the head, revealed right side sphenoid sinusitis, and left otomastoiditis. Blood tests showed activated lymphocytes in the peripheral smear, and positive IgM for cytomegalovirus (CMV). Patient maintained under Augmentin. Lumbar puncture revealing raised pressure (> 50 cm water). CSF studies show a

minimally raised serum protein level of 47 g/L, a glucose level within normal limits, gram stain was negative and revealed no bacterial organisms. Findings of cytology and other meningitis workup were negative. Differential diagnosis at that point included possible Pseudotumor cerebri or idiopathic intracranial hypertension. Venous access was obtained, and acetazolamide was administered, headache, vomiting and fever showed daily improvement, as well as partial regression strabismus and optic disc swelling. However, magnetic resonance imaging (MRI) performed 1 week after admission revealed, 30% occlusion of left transverse and sigmoid sinus in consideration of cerebral venous thrombosis, beside right mastoid inflammation process and sinusopathy. Immediately low-molecular-weight heparin was started. Further investigation revealed polymorphism of the hypofibrinolytic plasminogen activator inhibitor type 1 gene.

Discussion:

Cerebral venous sinus thrombosis (CVT) is a rare life-threatening disorder in childhood that is often misdiagnosed. The diagnosis of CVT somehow is difficult and can lead to a delay in treatment. The average from presentation to diagnosis is 1 week. Thus why, CVT should have a high clinical suspicion in patients who present with the risk factors. Imaging and lab workup can help in the differentiation between CVT and other similar diagnoses.

Most common ER radiological study is the head CT, CT without contrast relevant associated finding with CVT; however anywhere from 25- 45% of patients with CVT can have a normal head CT(6). Hypodensity of cortical vein or dural sinus are the most common finding in non-contrast head CT. MRI is more sensitive than a CT and should be performed in high suspicion of CVT and the head CT results were negative(7).

Routine blood work such as CBC, chemistry and coagulation profile should be performed to discover any underlying hematological disorder. A D-Dimer should be ordered to exclude suspicion of CVT, however D-Dimer not necessarily confirm a diagnosis of CVT. A lumbar puncture also may help as diagnostic and therapeutic value, CSF abnormal result are generally nonspecific for CVT(8). Prothrombotic abnormal conditions are the most common cause of CVT. Studies such as Antithrombin Protein C, Protein S, Factor V Leiden and lupus anticoagulant should also be performed. In pediatric population the most common causes of CVT are lymphoma, lupus, or leukemia and all these underlying conditions should be ruled out.

CVT management consists of treating the underlying cause first; however 10- 15% of patients has idiopathic causes, infectious require antibiotic treatment, or brain tumors may require surgical resection. The main corner of treatment for prothrombotic conditions is an anticoagulant such as LMWH, UFH, or warfarin(9). Many studies prefer heparin due to the short half-life if hemorrhage occur as adverse drug effect. Overall, treatment duration mainly depends on the underlying conditions, generally treatment take average, 6-12 months with INR goal of 2-3. Patients with prothrombotic disorders should be mentioned on anticoagulation for long life.

The last decade has been a period of rapid advances in the management of CVT using recanalization therapies by fibrinolytics or direct mechanical thrombectomy. Effectiveness of this method still under studying but there are small studies available. Recanalization therapy recommended for deteriorated patient despite adequate anticoagulation control or absolute contraindication to anticoagulation. There are only anecdotal reports of use of aspirin for cortical venous thrombosis. No randomized trial or even a case series exist on the use of antiplatelet agents(10).

CVT prognosis was very bad in the past, however recent studies show good outcome specially with new treatment modalities. Anticoagulation is the most widely used and accepted modality with favorable outcomes documented in two randomized controlled trials. CVT recurrence rate generally is low; however there are variable estimated risk, but in general consideration the recurrence rate is between 2.8-6.5% annually with adequate treatment(11).

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