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Case Report

Case Series of Two Patients Presenting with Cerebral Venous Sinus Thrombosis in Association with Ulcerative Colitis

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Abstract

Cerebral venous sinus thrombosis is a rare form of stroke of variable clinical presentation. Most cases are typically idiopathic but some cases are associated with other hypercoagulable disorders. Its association with inflammatory bowel diseases (Crohn's and ulcerative colitis) is rare, therefore presents significant diagnostic and therapeutic challenges. Because, this association has worse clinical outcome, therefore prompt recognition is mandatory. Any patients with the diagnosis of ulcerative colitis with neurological symptoms should urgently prompt the physician to suspect and diagnose CVST.

We are presenting two cases, the first case presented with neurological symptoms on presentation and later on ulcerative colitis was diagnosed based on her prior history while the second patient was known to have ulcerative colitis and presented with CVST. Both cases were managed with anticoagulation and made good neurological recovery as well as had remission of their intestinal symptoms.

Keywords: Cerebral venous sinus thrombosis; Ulcerative colitis; Anticoagulation

Introduction

Cerebral venous thrombosis (CVT) is a rare form of stroke, accounting for around 0.5% of all strokes[1], thatpresents with varied clinical manifestations mimicking several other neurological disorders and therefore it is very common to be misdiagnosed.

Cerebral sinus thrombosis has been reported as an uncommon complication of inflammatory bowel diseases like crohns and ulcerative colitis. The association of cvst with ulcerative colitis (UC), has been found in up to 7.5% of cases.[2] Therefore, diagnosis needs to be considered in any patient with IBD presenting with neurological symptoms. We are presenting two cases of UC presenting with cerebral venous sinus thrombosis.

Case 1

Our first patient is 20 years old Saudi female, who presented with headache and new onset generalized tonic clonic seizure (5 episodes in a day) three weeks after a full term normal vaginal delivery. There was history of vomiting and abdominal pain one week ago and was treated in a local hospital with IV hydration.

She was afebrile but looked very pale and dehydrated but neurological exam did not reveal any abnormality or focal neurological deficit. Her laboratory parameters revealed microcytic, hypochromic anemia with Hb of 7.1gm/dl, rest of the biochemistry and inflammatory parameters were normal. Considering her postpartum state and dehydration with history of seizures, provisional diagnosis of cerebral venous sinus thrombosis was entertained and her non enhanced axial computed tomography head revealed right occipital dense lesion at the area of the right transverse sinus and CT venogram showed filling defect in left internal jugular vein, superior sagittal sinus and right transverse sinus (Figure 1 (A and B)).

On systemic review, she informed about repeated admissions during the pregnancy with abdominal pain, vomiting and loose stools with fresh blood at some occasions. She received blood transfusions twice and treated with antibiotics but no workup was done and most of her symptoms were attributed to pregnancy, therefore gastroenterology evaluation was sought and based on her symptoms, the provisional diagnosis of inflammatory bowel disease was considered that was confirmed by colonoscopy and CT enterography with the findings suggestive of severe ulcerative colitis (Figure 1C).

Patient was managed with IV hydration, antiepileptic medications and IV heparin followed by low molecular weight heparin (Enoxaparin) and for UC, she was started on Adalimumab. During her hospital stay, she improved with no further seizures as well as her abdominal symptoms also improved. Hypercoagulable work up showed mildly positive lupus anticoagulant antibodies. Unfortunately, the patient lost follow up after discharge.

Case 2

Case 2 is a 15-year-old boy with a 3-year history of distal ulcerative colitis (UC). Diagnosed by progressively worsening

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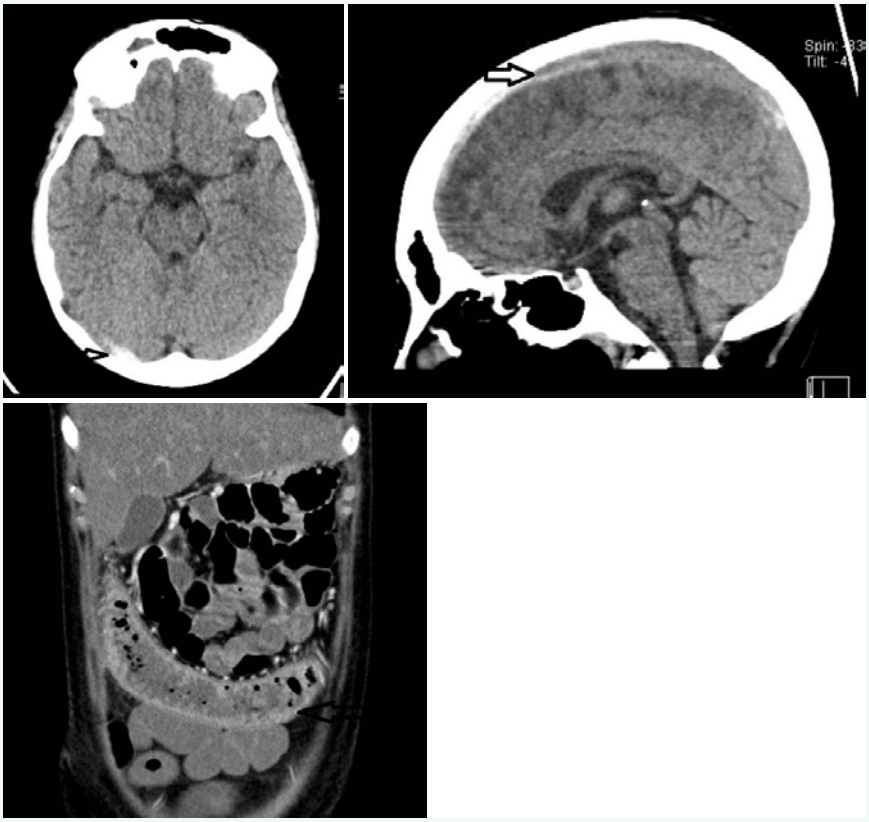

Figure 1 (A) plain computed tomography of the brain indicating filling defect in Right transverse sinus. (Arrow)
(B) Plain computed tomography of the brain indicating superior sagittal sinus thrombosis (arrow)
(C) CT enterography showing colonic wall thickness with mucosal enhancement and loss of haustrations (Arrow).

symptoms of abdominal pain, urgency, diarrhea and rectal bleeding outside of our hospital that was confirmed by CT abdomen and colonoscopy, started on therapy consisting of oral corticosteroids and mesalamine and azathioprine, but being noncompliant and altering medication by him. He subsequently presented to the emergency department with acute onset of headache and severe neck pain with low grade fever being attributed to recent sore throat. He was admitted one month ago under gastroenterology services with the history of abdominal pain and bloody diarrhea and again one week back with the similar complaints and was treated with ciprofloxacin and steroid was tapered. His neurological examination including fundoscopy was unremarkable. With the background history of severe ulcerative colitis, now presenting severe headache, the diagnosis of CVST was suspected and CT and CT venogram head was arranged. Plain head CT head was unremarkable and CT venogram showed an acute sagittal sinus, left transverse and sigmoid sinus thrombosis (See Figure 2 A,B). As he was febrile, so after excluding the contraindications, CSF analysis was performed that was negative for any CNS infection. Patient was started on therapeutic doses of enoxaparin and good hydration. Gastroenterologists were contacted and he was restarted on oral steroids and mesalamine.

He was anemic and required blood transfusion initially but later on his hemoglobin remained stable. Prothrombotic screen (prothrombotic screen included protein C, protein S and antithrombin III deficiency, Prothrombin gene mutation, factor V Leiden mutation, lupus anticoagulant, anticardiolipin antibody and fasting homocysteine) was negative. His colitis improved as well as his neurological symptoms improved completely within a week and he was started on oral anticoagulation and discharged home in a stable condition.

Discussion

As highlighted by our two cases, presenting features in cvst can be highly variable, ranging from headache or neck pain to seizures, particularly if the bowel disease is active.

Cerebral venous thrombosis (CVT) is a rare type of stroke that can happen at any age, including the neonates, and it is responsible for approximately 0.5% of all strokes [1].

Clinical presentations are variable and can range from simple headache to severe neurologic dysfunction depending on the degree of intracranial pressure increases, the locations of the sinuses affected and the resultant venous infarction or hemorrhage [2]. Most cases of CVT in young people are usually idiopathic, hence some reports have pointed out the existence of an underlying disorder [3] the association among CVT and UC vary remarkably worldwide. In certain parts of the world especially north America and Europe, UC is supposed to be the

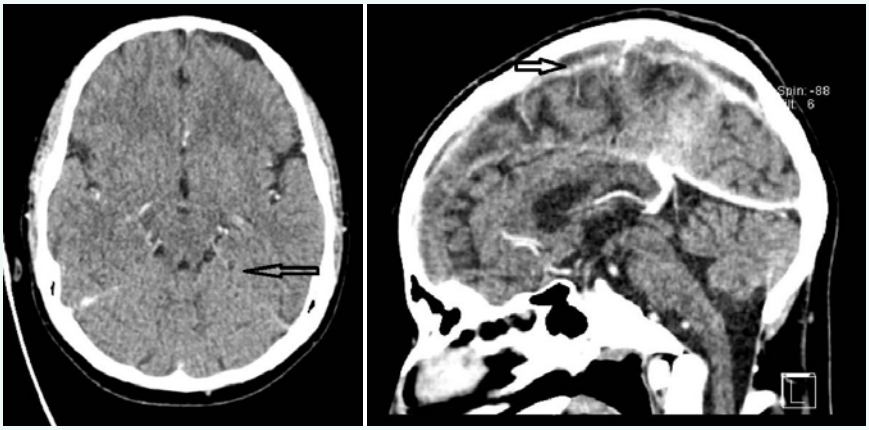

Figure 2 (A) Post-contrast computed tomography of the brain indicating filling defect in left transverse sinus. (Arrow) (B) Post-contrast computed tomography of the brain indicating superior sagittal sinus thrombosis (arrow)

main cause of cerebral venous sinus thrombosis [4], while it is considered to be a rare cause in Japan and according to study published in 2013 by Ikeda et al., fewer than 20 cases of cvst with UC have been reported have been reported from there [5].

Ulcerative colitis is one of the two entities of chronic inflammatory bowel disease which affects the lining of the large intestine (mainly colon) and rectum. The etiology of UC is idiopathic but is considered to be the result of complex interaction between environmental factors in genetically susceptible people [6] with the awareness about the disease and availability of the diagnostic modalities, recently more cases have been identified with this disorder [7,8].

Any age group might be affected by UC, but usually there are two peaks at ages 15-30 and then at 50-70. Extraintestinal manifestations are reported in almost 40% of the adult patients with UC involving the bones, joints, skin, lungs, blood, kidneys, eyes, liver and peripheral as well as central nervous system [9]. Thrombosis is reported in almost 6.5% of the patients with active IBD [10], the incidence of CVST being more common with UC than crohns disease (almost 7.5%) [2].

Patients with inflammatory bowel disease are at risk of having hypercoagulable state and therefore, the risk of various thrombotic events (pulmonary, VT as well as venous sinus thrombosis) is higher and usually affects patients at younger age in comparison to the patients without IBD [11]. The risk of thrombosis is more frequent during the IBD relapse but the cases have been reported even 10 years after colectomy for UC [12].

The outcome of CVT is largely unpredictable, and consensus about treatment has not been reached. In general, Systemic anticoagulation is considered to be the primary therapy for CVT even in individuals presenting with hemorrhagic infarcts [13,14]. But in patients who have cvst complicated by IBD, there is a still controversy about the treatment as many physians believe that thromboembolic events subside naturally and AC can be potentially dangerous because of mucosal bleeding from the UC lesions and patients are given only antiepileptic drugs [15,16].

However, our both cases were treated with systemic anticoagulation despite first patient having severe ulcerative colitis and fresh bleeding per rectum but both showed good neurological outcome as well as improvement in gastrointestinal symptoms.

Conclusion

Patients with UC who suffer from cerebral venous thrombosis can be safely and effectively treated with anticoagulation and have a good outcome with early recognition and management; hence it's very important to suspect CVST in any patient with UC who have even subtle neurological symptoms.

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