DOI: 10.7860/JCDR/2016/20105.7883

Case Report

Internal Medicine Section

Unusual Case of Cerebral Venous Sinus Thrombosis in Patient with Ulcerative Colitis in Remission

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ABSTRACT

Ulcerative colitis (UC) is an idiopathic autoimmune inflammatory disease of the gastrointestinal tract. Cerebral venous sinus thrombosis along with deep vein thrombosis, pulmonary embolism and arterial thrombosis have occasionally been reported as a complication in the active phase of UC being attributed to its pro-thrombotic state. This paper depicts a 38-year-old female with a history of UC in remission who developed sudden onset headache, blurring of vision and seizures. Subsequent diagnosis of cerebral venous sinus thrombosis was made with MRI venography and treated with low molecular weight heparin with complete resolution of symptoms. The highlights of this case underscore the importance of evaluating cerebral venous sinus thrombosis as a cause of acute onset neurological deterioration in a setting of inflammatory bowel disease. It also emphasizes on the hypothesis that the risk of venous thrombosis or other hypercoagulable states have no direct relationship with the disease activity or flare-up.

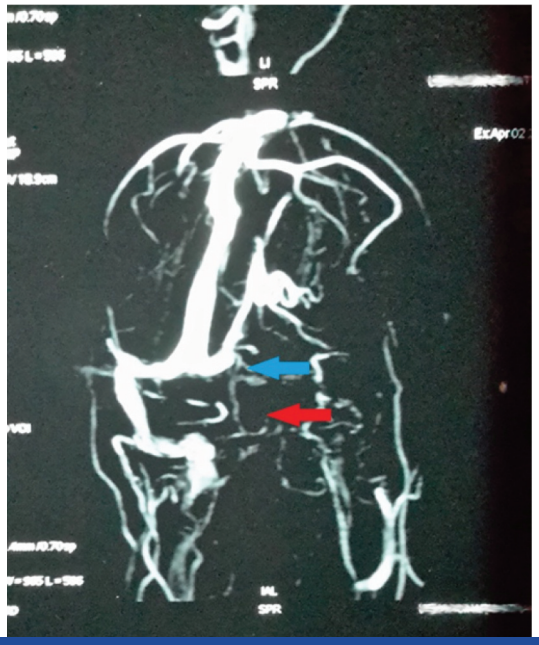
Keywords: Disease activity index, Inflammatory bowel disease, Partial mayo score

CASE REPORT

A 38-year-old female presented to the emergency department of MKCG Medical College and hospital with chief complaints of a two day history of generalized headache associated with nausea, vomiting, blurring of vision and photophobia. This was associated with one episode of generalized tonic-clonic seizure a few hours prior to arrival at the ER. Her previous history was not suggestive of any prior episode of transient ischemic attacks, stroke or seizures. She was diagnosed as a case of Ulcerative Colitis (UC) two years back. She was on oral steroids, 5-Amino Salicylic Acid (5-ASA) and sulfasalazine following which she was symptom-free for last one year.

On examination she was afebrile and normotensive. Glasgow Coma Scale (GCS) was 11/15 (E3V4M4) with bilateral plantar extensor. Fundoscopy revealed changes suggestive of early papilloedema in both eyes. Routine tests revealed a normocytic normochromic anaemia with a haemoglobin of 9.8 g%, total leukocyte count of 10,400/mm^3, Erythrocyte Sedimentation Rate (ESR) of 18mm in 1st hour, platelet count was 3.01 lakhs/mm^3. C-Reactive Protein (CRP)was 10.6 mg/dl. Prothrombin Time (PT) was 13.0 seconds (Reference range: 12-13 seconds), International Normalised Ratio (INR) was 1.45, activated partial thromboplastin time (aPTT) was 36 seconds (Reference range: 32-40 seconds). Liver and renal function tests along with serum electrolytes were within normal limits. Sickling test was negative. Blood and stool cultures collected on the day of admission were negative for known pathogens. Stool routine and microscopy revealed few leukocytes and erythrocytes. Rectal examination was negative for melena, bleeding or diarrhoea. Non-Contrast Computerised Tomography (NCCT) scan of the head was normal. Owing to her young age and absence of known risk factors for stroke, cerebral venous sinus thrombosis was suspected and MRI venography was ordered which showed a filling defect in left transverse sinus and left sigmoid sinus [Table/Fig-1]. A provisional diagnosis of Cerebral Venous Sinus Thrombosis (CVST) was made and she was immediately put on intravenous Low Molecular Weight Heparin (LMWH) therapy and phenytoin.

Prothrombotic work-up and coagulation studies were negative for protein C and protein S activity, Antithrombin III activity, Anti-phospholipid antibody, Homocysteine and factor V Leiden mutation. The Ulcerative Colitis Disease Activity Index (UCDAI) was calculated using partial Mayo score [1,2]. The score was 0 which clearly indicated that the disease was in remission phase and there was no flare-up of the disease process whatsoever. She was discharged after 10 days with oral warfarin and oral phenytoin with almost complete resolution of symptoms and no seizures since the day of admission. She was symptom-free till last follow-up i.e. at 4 months of discharge from the hospital.



[Table/Fig-1]: Magnetic Resonance Venography showing non visualization of left transverse (blue arrow) and left sigmoid sinus (red arrow), right transverse sinus and right sigmoid sinus are clearly visible.

DISCUSSION

Ulcerative Colitis (UC) is a form of Inflammatory Bowel Disease (IBD) that primarily causes inflammation and ulcers in the colon and rectum. Many hypothesis and theories have been postulated to explain how exactly UC contributes to a hypercoagulable state. Cerebral Venous Sinus Thrombosis (CVST) is a rare but potentially disabling extra-intestinal complication of IBD. The yearly

incidence of CVST in IBD is reported to be around 1.3% - 6.4% [3]. Thromboembolic phenomena affects vessels of both arterial and venous system. The frequency of CVST appears to be slightly greater in patients with UC as compared with Crohn's disease [4]. The superior sagittal sinus and lateral sinus are more commonly involved [4]. Headache, seizures, dysphasia, blurring of vision, focal neurological deficits are often the initial set of symptoms at presentation.

The mechanisms and pathophysiology involved in CVST is still a matter of debate. Prothrombotic states such as increased fibrinogen levels, hyperhomocysteinaemia, Antithrombin III deficiency, Protein C and S deficiency, thrombocytosis, presence of anti-cardiolipin antibodies, increased factor V and VIII levels have been proposed as possible explanations [5]. Alteration in plasma and mucosal haemostasis such as increased endothelial activation, impaired inhibition of coagulation and various other inflammatory changes associated with the disease process such as release of acute phase reactants, activation of coagulation cascade shifts the balance towards a pro-coagulative state [6].

The usual treatment strategies include intravenous LMWH therapy followed by oral anticoagulation with warfarin when the patient is relatively stable [7]. Mechanical thrombectomy have been employed in a few cases. Drug interactions used in the treatment of baseline IBD such as mercaptopurine and azathioprine further complicates the course of treatment and poses a significant problem in achieving therapeutic anticoagulation [8,9].

The association of disease activity of IBD and incidence of thromboembolic phenomena is still controversial. Kristensen et al., reported a significant direct association of disease activity of IBD with increased risk of morbidity and mortality [10] whereas Katsanos et al., reported the presence of CVST in active disease in 78.4% and in quiescent disease in the rest 21.6% [11]. Till date only a handful of case reports have been published [12,13] and the dilemma in management continues to haunt researchers and physicians alike.

CONCLUSION

In conclusion, our case highlights the onset of CVST in the quiescent or remission phase of the disease. Any sudden onset and or acute worsening of neurological status such as headache, mental confusion, convulsions and papilloedema on fundoscopy in a patient with IBD should alert the treating physician about the possibility of CVST so that timely intervention could be employed to prevent the devastating complications of this fatal and disabling condition.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Mar 12, 2016**
Date of Peer Review: **Apr 16, 2016**
Date of Acceptance: **Apr 19, 2016**
Date of Publishing: **May 01, 2016**