

Cerebral sinus thrombosis: a rare but fatal complication of inflammatory bowel disease

Abstract

Inflammatory bowel disease (IBD) is an idiopathic autoimmune disease. Extra intestinal manifestation of IBD has been reported to be 25 to 35%. There is a well-known risk of thrombosis in patients with IBD. It rarely involves cerebral vasculature in the form of arterial stroke or venous sinus thrombosis. This case is about 35 yrs old male who was a known case of ulcerative colitis for the last 10 years. He presented with history of severe thunderclap headache for the last few hours along with vomiting. He also had three episodes of seizure, generalized tonic clonic in nature. The examination does not show any focal asymmetry except mild drowsiness. His CT brain showed Focal cortical and subcortical area in the left posterior parietal region displaying multiple foci of hyper density with perifocal edema. CT venogram showed an intraluminal filling defect in the superior sagittal sinus suggestive of venous sinus thrombosis. Conclusion: Cerebral venous sinus thrombosis are considered as one of the rare but fatal complication of inflammatory bowel disease. It must be considered in all those with inflammatory bowel disease wherein absence of any other vascular risk factor patient presents with acute severe headache and focal or diffuse neurological symptoms including seizure.

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Introduction

Inflammatory bowel disease (IBD) is an idiopathic disease caused by a deregulated immune response to host intestinal micro flora. The two major types of inflammatory bowel disease are ulcerative colitis (UC), which is limited to the colon, and Crohn disease (CD), which can involve any segment of the gastrointestinal tract from the mouth to the anus in the form of "skip lesions," and is transmural. Extra intestinal manifestation of IBD has been reported to be 25 to 35%.¹ Inflammatory bowel diseases (IBD) are associated with the occurrence of thrombotic complications. There is a well-known risk of thrombosis in patients with IBD with an overall incidence of 1.3-7.5%. It rarely involves cerebral vasculature in the form of arterial stroke or venous sinus thrombosis.² There are very few cases of Crohn disease associated with cerebral sinus thrombosis during disease activity.³

Case history

This case is about 35 yrs old male who was a known case of ulcerative colitis for the last 10 years and was on regular medication including sulfasalazine. For the last one and half years, he was not taking sulfasalazine regularly and had completely stopped taking these 2 months before. Two weeks prior to this presentation, he developed diffuse abdominal pain, mild fever and episodes of diarrhea (4 to 5 episodes per day). He went to medical physician who started him topical mesalazine and corticosteroids preparation along with advice of continuous oral medication for his inflammatory bowel disease but patient refused to restart oral medication. He was treated for acute episodes and discharged.

Two weeks later, he presented to the emergency dept. with history of severe headache for the last few hours along with vomiting. This headache was sudden in onset and was thunderclap in nature, which was worsening in nature. He was brought immediately to the emergency department. He started having seizure, generalized tonic clonic in nature with up rolling of eyes, generalized jerking of limbs tongue bite and frothing of mouth followed by confusional state and restlessness. He had three episodes within 2 to 3 hours. There was no history of any fever, loss of consciousness, any visual and bulbar

symptoms. There was no history of any head injury, trauma and accident. Family history was unremarkable.

Examination at the time of admission showed normal temperature along with blood pressure 145/90. His Glasgow Coma Scale was 15/15. He was conscious, but drowsy. There was no pupillary abnormality. Sizes were 2 mm and were reactive to light. There was no extra ocular abnormality. No facial abnormality was observed during examination. In the motor system examination, the tone was bilateral normal. Reflexes were generalized brisk. The power was 5/5 in all muscle groups bilaterally and the planter was down going. Sensory examination was unremarkable. There were no cerebellar signs. There was a mild neck rigidity and kerning signs was positive. Systemic examination was unremarkable.

CT brain was done immediately in an emergency, which showed Focal cortical and subcortical area in the left posterior parietal region displaying multiple foci of hyper density with perifocal edema. CT brain with contrast, showed no enhancement of the previous described area, at the cortical and subcortical left posterior parietal region. The picture was suggestive of possibility of hemorrhagic infarction could be due to venous sinus thrombosis. CT Venogram was done immediately, which showed intraluminal-filling defect in the superior sagittal sinus suggestive of venous sinus thrombosis.

His blood work up showed WBC 13000 (4000 -11000), Hb 10.1 (11-15 g/dl) with microcytic hypochromic picture. Rests of the blood counts were normal. His liver function, renal function test, serum electrolyte, blood sugar fasting and coagulation profile were normal. In vasculitic profile anti cardiolipin IgG 3.8 U/ml (negative <10 U/ml), anticardiolipin IgM 1.3 U/ml (negative <10), anti-DS DNA antibodies <10 (<100 IU/ml Negative), Anti-Nuclear Factor negative, c-ANCA - PR3 0.7 Units (negative <5 U/ml) p-ANCA - MPO 0.9 Units (negative <5 U/ml) Anti-THRM III 80% (66-124), Protein C 72% (54-166) and Protein S 58% (54-103) His ESR was 50 mm/1hr (0-20) and C reactive protein was 45 mg/l (0.3-5) suggestive of active inflammatory process. He has been offered to go for a colonoscopy and biopsy. However, as he was already diagnosed case of ulcerative colitis and he refused to have new intervention so was not done.

The patient was immediately stabilized in an emergency. He was started on infusion phenytoin 1 gram as bolus. After establishing the diagnosis, he was shifted to the medical intensive care unit. He was started on low molecular weight heparin according to weight along with oral anticoagulants. He had been given symptomatic care for headache and vomiting. The patient started improving. Gastroenterology department was consulted to look for ulcerative colitis. The patient was not taking any medication for Ulcerative colitis (UC). He was started on mesalazine 500mg twice a day along with some topical preparation for inflammatory bowel disease.

Discussion

Our patient has a long-standing history of ulcerative colitis and was well maintained on sulfasalazine but for the last few months, he was not on any medication for UC (Ulcerative colitis). Moreover, he had a relapse of bloody diarrhea few weeks ago. His clinical and laboratory data were favoring active inflammatory disease. Personal and family medical history did not reveal any other prothrombotic risk factors, or a bleeding diathesis. Therefore, this sinus thrombosis were thought to be related to the activity of the disease, especially in the absence of any other risk factors.

Table 1 Case reports showing sinus venous thrombosis in patients with inflammatory bowel disease

Reference	Age/Sex	Inflammatory bowel disease (IBD)	Treatment	Final Outcome
Philips et al. ⁸	35/Female	Ulcerative colitis	Systemic anticoagulation	Complete recovery
Kao et al. ⁹	14/ Female	Ulcerative colitis.	Systemic anticoagulation	Partial recovery
Tsujikawa et al. ¹⁰	27/ Male	Ulcerative colitis	Systemic anticoagulation	Complete recovery
Kao et al. ⁹	20/ Female	Ulcerative colitis	Systemic anticoagulation	Complete recovery
Wasay et al. ¹¹	23/ Female	Ulcerative colitis	Systemic anticoagulation	Partial recovery
Samal et al. ¹²	20/Male	Crohn's disease	Oral anticoagulants	Partial resolution of symptoms
Srivastava et al. ¹³	29/ Male	Ulcerative colitis	LMWH followed by systemic anticoagulation	Complete recovery

So if the patient is presenting with recent unusual headache, stroke like symptoms, seizures, or any other brain syndrome, due to its highly variable presentation, we should consider CVST (cerebral venous sinus thrombosis) as one of the top most differential and this suspicion should be even stronger in a young person with neurological signs in the presence of active inflammatory bowel disease and absence of any other known risk factors for venous thrombosis.

Headache is considered as one of the most frequent signs of cerebral venous thrombosis, which gradually increases over a period of days. It can also present as an abrupt thunderclap headache like that of a patient with subarachnoid hemorrhage.¹⁴ In our patient headache was also very prominent and most important symptom. Imaging also showed evidence of mild subarachnoid hemorrhage as well. Seizure is also very common in patient with CVT, found in about 40% of the patients with CVT. In most of the patient bilateral symptoms are found, whereas unilateral focal neurological signs are less common in patients with venous thrombosis.

After clinical evaluation for cerebral venous thrombosis, the next step is to go for MRI brain along with the MRV (venography). However, if MRI facilities are not available CT Venography is recommended as an alternative. If we look at the overall accuracy of head CT combined with CT venography, it is 90 to 100 percent,

Many central and peripheral neurological disorders have been reported in patients with IBD. These include peripheral neuropathies, myopathies, focal central nervous system defects, convulsions, confusional episodes, meningitis, syncope, optic neuritis, and sensorineural loss.⁴ Most of these symptoms are related to underlying autoimmune pathophysiology.

The cause of hyper coagulation and thromboembolism in inflammatory bowel disease has not been yet fully understood until now, but coagulation factor abnormalities such as elevated fibrinogen level, factor V, factor VIII, an increase in circulating thrombin-ant thrombin complexes, and decreased ant-thrombin III have been mentioned in literature. In addition to that thrombocytosis and increased platelet aggregation have also been documented.⁵⁻⁷ However, there is no substantial evidence to correlate hematological and coagulation abnormalities with cerebral sinus thrombosis. But there are many other similar cases have been reported in the literature where there is a clear association of IBD in the form of Ulcerative colitis or Crohn's Disease (Table 1).

(Insert Table 1 Here)

depending on the occlusion site.¹⁵ Guidelines from the AHA/ASA published in 2011 consider CT venography to be at least equivalent to MR venography in the diagnosis of CVT.¹⁶

After confirmation of the diagnosis, anticoagulant therapy is recommended to promote spontaneous thrombus resolution prevent Thromboembolism.¹⁷ For adults with symptomatic CVT, with or without hemorrhagic venous infarction, it is recommended to start initial anticoagulation therapy with subcutaneous low molecular weight heparin (LMWH) or intravenous heparin followed by oral anticoagulation according to the clinical condition of the patients.¹⁸

Conclusion

Cerebral venous sinus thrombosis are considered as one of the rare but fatal complication of inflammatory bowel disease. It must be considered in all those with inflammatory bowel disease wherein absence of any other risk factor, the patient presents with acute severe headache and focal or diffuse neurological symptoms including seizure. With timely diagnosis and proper treatment, we can reduce the morbidity and mortality of the patient. In addition to that, the treatment of the underlying active IBD should be continued to control the relapses as occurred in our patient (Figure 1).

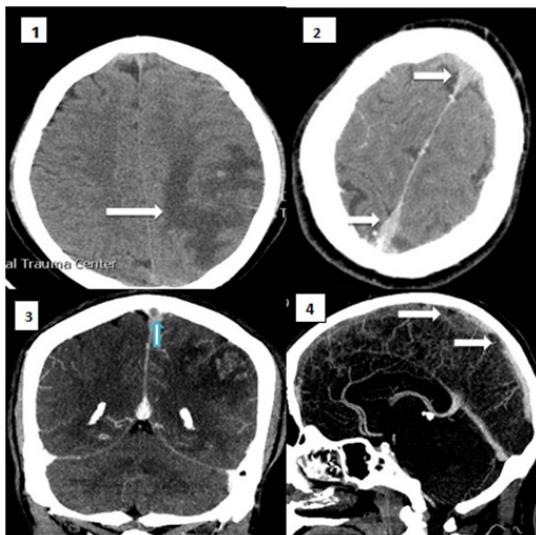


Figure 1 (1) Focal cortical and subcortical area seen at the left posterior parietal region displaying multiple foci of hyper density with perifocal edema. (2) Superior sagittal sinus thrombosis. (3,4) CT angiography showed superior sagittal sinus thrombosis.

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None.

Conflicts of interest

None.

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