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Crohn's Disease Presented with Cerebral Venous Sinus Thrombosis: A Case Report

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Abstract

Cerebral venous sinus thrombosis (CVST) is a fatal condition if not treated accordingly. As it has a wide variety of presentations and association with many other diseases, often we felt diagnostic dilemmas and challenges. This consumes our time as well as put a bad impact on patient's recovery. As like many other inflammatory conditions, Inflammatory Bowel disease (IBD) has also association with venous thromboembolism (VTE). Usually VTE's are associated with the flare of IBD. The common sites are deep veins of legs, pulmonary veins, portal veins and mesenteric veins. Although not so common still there are some case reports regarding CVST in Crohn's disease (CD) patients like ours one. A 31 years old lady diagnosed as a case of Crohn's disease 1 year earlier presented with headache, vomiting, left sided hemiparesis and several episodes of convulsion. Magnetic resonance imaging (MRI) of the brain with venography showed thrombosis of superior sagittal sinus. Then she was immediately anticoagulated with low Molecular weight heparin (LMWH) followed by Rivaroxaban 20mg daily. *[Journal of National Institute of Neurosciences Bangladesh, January 2025;11(1):92-95]*

Keywords: Inflammatory bowel diseases; Crohn's disease; cerebral venous sinus thrombosis; Anticoagulation

Introduction

Cerebral Venous Sinus Thrombosis (CVST) is a rare form of stroke caused by thrombosis (blood clot) in the dural venous sinuses, which drain blood from the brain[1]. Unlike arterial strokes, Cerebral venous sinus thrombosis affects the venous system and can lead to increased intracranial pressure, cerebral edema, venous infarction, or hemorrhage.

Cerebral venous sinus thrombosis can present with a wide range of symptoms, making diagnosis challenging[2]. Common clinical features include headache (often the earliest and most frequent symptom), seizures, focal neurological deficits, altered consciousness, and signs of raised intracranial pressure such as papilledema and vomiting. It occurs in all age groups and both sexes but is more common in young adults, particularly females, due to risk factors like pregnancy, postpartum state, oral contraceptive use, and thrombophilia. Other risk factors include infections, malignancies, dehydration, autoimmune diseases, and trauma. Because of it's wide variety of presentations, the diagnosis is often missed & overlooked without searching provocating factors meticulously. And a timely diagnosis is very crucial, as it has a high fatality rate if not treated accordingly[3].

Inflammatory bowel disease (IBD) is a group of chronic, relapsing inflammatory conditions of the gastrointestinal (GI) tract, primarily including Ulcerative Colitis (UC) and Crohn's Disease (CD). These conditions are characterized by inappropriate and ongoing inflammation of the intestinal mucosa, likely driven by a dysregulated

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immune response to intestinal microbes in genetically susceptible individuals. While Ulcerative Colitis is limited to the colon and rectum with continuous mucosal inflammation, Crohn's Disease can affect any part of the GI tract from mouth to anus and typically shows transmural and patchy inflammation. Common symptoms include abdominal pain, diarrhea (often bloody in UC), weight loss, fatigue, and extra-intestinal manifestations like arthritis, uveitis, and skin lesions.

Patients with IBD are prone to VTE, particularly in the active stage of the disease[2]. The common sites are deep veins of legs, pulmonary veins, portal veins and mesenteric veins[3]. Although involvement of cerebral vein & sinuses is uncommon, still it may happen[2]. Among inflammatory bowel disease, Ulcerative colitis (UC) is more prone to it than CD[2]. Here, we describe a case of cerebral venous thrombosis as a complication of active CD.

Case Presentation

A 31 years old married, regularly menstruating lady having two children with having history of two early fetal loss presented with sudden onset severe dull aching global headache followed by a sudden onset rapidly progressive left sided spastic hemiparesis which become static over a few hours & confined her to bed. She presented to us within 24 hours of symptom onset. During her course of this illness, she encountered four episodes of Generalized Tonic-Clonic Seizure (GTCS), each episode persists about 1-2 minutes with prominent post-ictal confusion. There was no diplopia, visual obscuration or blurring of vision, paresthesiae, difficulty in swallowing or in speech. Her fundoscopic examination reveals a normal appearing disc & periphery of retina. Her bladder habit remains normal. On systemic query we found no history of arthritis or arthralgia, skin rash, oral ulceration or red eye. On top of this, she was a diagnosed case of Crohn's disease on the basis of symptomatology & some rational supportive investigations. About a year ago, she developed occasional colicky abdominal pain particularly on mid of her abdomen followed purging of loose unformed stool approximately 4-5 times per day. The stool remains yellow in color, occasionally mixed with mucus but there was no foul smelling & the stool was not greasy. There was no history of weight loss, vomiting, passage of blood mixed or black tarry stool. On this clinical context with a high fecal calprotectin she was undergone colonoscopy followed by biopsy. Colonoscopy reveal multiple variable size ulceration on ileum near ileo-caecal valve (Figure I) and on biopsy from this ileal ulcer reveals lympho-plasmo-histocytic cell infiltration on lamina propria manifesting chronic active ileitis with lymphoid hyperplasia with no obvious granuloma. MR enterography showing narrowing &, wall enhancement at proximal jejunal & distal ileal region (Figure II). Her IGRA was negative, MT reveal 3 mm and Genexpert ultra of ileal sample fails to detect any MTBC. After all this workup her gastroenterologist started prednisolone 1mg/kg/day for 3 weeks and then tapper over next 3 months. She became quite well 1-2 months followed by this. But again another exacerbation occurs & her gastroenterologist again put her on same drug with same regimen. Without significant improvement, however she developed these current acute conditions and presented to us. With prompt workup we establish her this episode

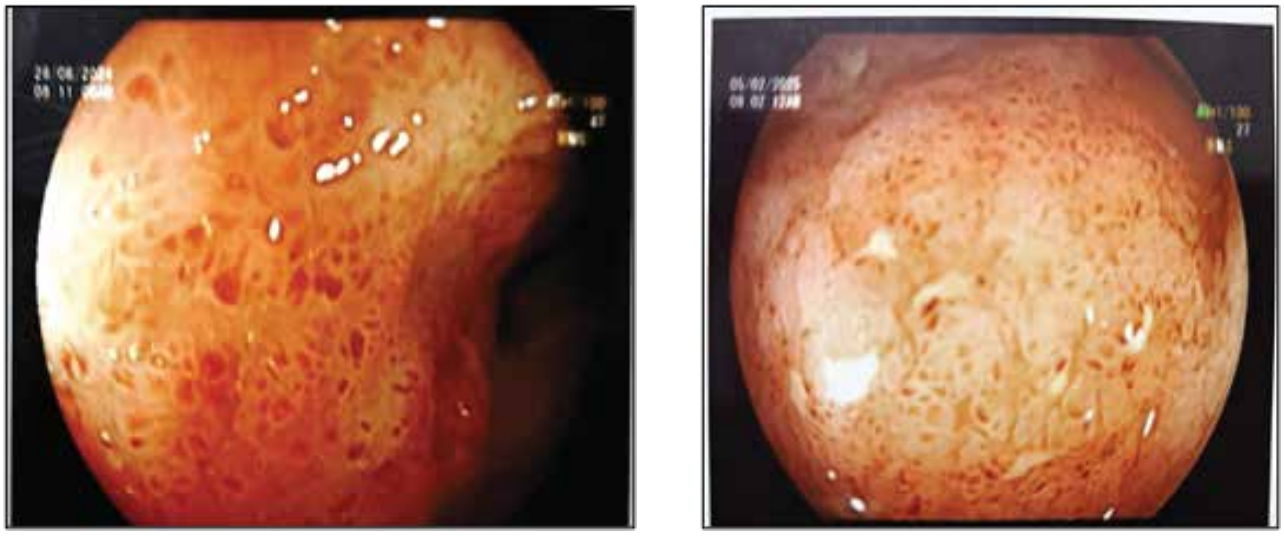

Figure I: Multiple ulcer of variable size seen at the terminal ileum with loss of normal vascular pattern

as a case of cerebral venous sinus thrombosis on the basis of luminal & marginal narrowing of Superior sagittal sinus as well filling defect at mid part of superior sagittal sinus on MR Venogram (Figure III). Concomitantly her d-dimer was high (3.96 mcg/ml). Blood tests for homocystine, ANA, APS panel, ANCA's albumin, CRP, thrombophilia screening, renal & liver function tests remain normal. After establishing a diagnosis of CVST, we started LMWH subcutaneously followed by oral rivaroxaban 20mg daily. During hospitalization to our Department her CDAI (Crohn's "Disease Activity Index) was initially 230 & after a week it became 180.

Figure II: MRI enterography features suggestive of enhancing wall thickening at proximal part of jejunum & distal ilium resulting luminal narrowing without any features of obstruction.

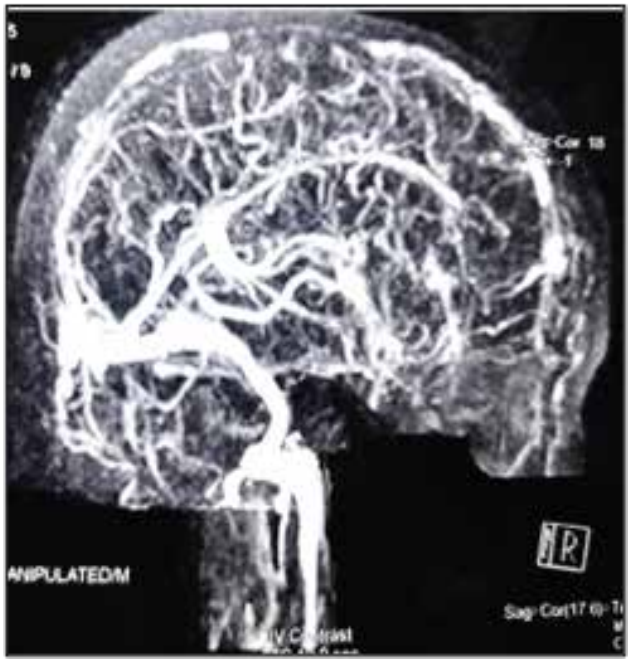

Figure III: MR Venography with Contrast showing narrowing, marginal irregularity & filling defect noted at superior sagittal sinus with cortical venous accentuation

Discussion

Cerebral venous sinus thrombosis is a venous stroke, rarer than arterial stroke and provoked by multiple conditions[4]. Presenting symptoms of cerebral venous sinus thrombosis can be due to increased intracranial pressure or focal parenchymal injury, with or without mass effect[5]. Among presenting symptoms headache is the most prevalent one (90.0%). Other includes signs and symptoms related to intracranial pressure include nausea, transient visual obscurations or vision loss (13.0% to 27.0%), papilledema, and diplopia (6.0% to 14.0%), cranial neuropathies (6.0% to 11.0%), seizure at time of presentation (20.0% to 40.0%), focal neurological deficits (20.0% to 50.0%), Coma&encephalopathy about 20% cases[5].

Symptoms tend to occur more insidiously than in other stroke types, and the majority will present more than 48 hours after onset. A minority may have more acute presentations with thunderclap headache or subarachnoid hemorrhage (less than 5.0%) or acute onset of focal neurologic deficits (5.0% to 40.0%)[5]. The clinical presentation of cerebral venous sinus thrombosis is varied and this creates a diagnostic challenge for clinicians.

Inflammatory bowel diseases can be complicated by a variety of extra-intestinal conditions, including venous thrombosis due to hypercoagulability. In cases of venous thrombosis, cerebral vascular involvement is very rarely observed[2-3]. It has been estimated that 1.3% to 6.4% of patients with inflammatory bowel diseases (IBD) are complicated by cerebral venous thrombosis (CVT) at some point of time during the course of their disease[2]. In a study that retrospectively analyzed data from 65 case reports of IBD patients with cerebral venous thrombosis, the average patient age was 29, and women experienced complications more frequently than men (37 vs. 28)[2]. The incidence of UC was almost double compared with CD (42 vs. 21), and active disease was reported in 78.4% of the cases[2]. The most common site of cerebral venous thrombosis was the superior sagittal sinus (50.7%), and thrombosis in multiple sites occurred in about 50% of cases[2]. The main risk factors for hypercoagulability were anemia (49%) and thrombocytosis (26.0%); however, there were many cases (23%) accompanied by coagulation abnormalities of fibrinogen, elevated factor

VII, elevated factor VIII, protein C deficiency, and protein S deficiency. In our case, cerebral venous thrombosis complicated during flare up of CD.

In patients with IBD complicated by cerebral venous thrombosis, favorable outcomes are possible with early diagnosis and appropriate treatment. However, when patients are not treated, the mortality rate can be as high as 50.0%[2]. Therefore, physicians should be especially careful to consider the possibility of cerebral venous thrombosis in patients with IBD, especially when there is active disease, as in our case. Treatment of cerebral venous thrombosis must continue for at least 3 months, but if there are risk factors for thrombosis, coagulation abnormalities, or thrombogenic mutation, then long term treatment of more than 6 months should be maintained[6]. Since the underlying IBD can be a cause of thrombosis, treatment of the inflammation itself is essential[7]. Thus, while treating cerebral venous sinus thrombosis with anticoagulation, we shouldn't forget to treat the primary disease that leading to this fatal complication.

Conclusion

Cerebral venous sinus thrombosis is frequently missed due to its wide spectrum of clinical presentation that might be confused with other pathologies. Thus, a high degree of suspicion is necessary for early detection and implementation of appropriate therapy, thereby reduce mortality and morbidity. IBD can cause VTE. Though Cerebral veins & sinuses are not a common site, still in appropriate clinical context, a suspicion of cerebral venous sinus thrombosis with IBD is rational.

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Contribution to authors:
Khaled Mosaraf, has conceived and designed the case report, analyzed the data & correlate them as per literature review, and wrote up the draft manuscript. Dipta Roy, Muhammad Saiful Islam, Chowdhury Neamul Hassan Refayet, Sk. Rezaul Haque, Ruhul Quddus, Md. Bakhtiar Azam &Mahmudul Islam involved in the manuscript review and editing. All authors read and approved the final manuscript.

Data Availability
Any inquiries regarding snd are available from the corresponding author on reasonable request.

Ethics Approval and Consent to Participate
Consent form was taken from the legal guardian of patient.

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