Cerebral venous sinus thrombosis in Crohn's disease

The empty delta sign

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ABSTRACT

يعد مرض الأمعاء الالتهابي من الأمراض الالتهابية المجهولة السبب التي تصيب القناة الهضمية، كما يُعرف ارتباط هذا المرض بخطر الإصابة بالتجلط الدموي حيث تتراوح نسبة الإصابة ما بين 1.3–7.5%. يعد حدوث التجلط الوريدي الجيبي المخي بعد الإصابة بمرض الأمعاء الالتهابي من المضاعفات غير الشائعة. ويُظن أن حدوث ذلك قد يكون نتيجة لتأهب وراثي وزيادة القابلية للتجلط أثناء نشاط الالتهاب المعوي. لقد تطرق الأدب الطبي لحالات من مرض كرون التي ترافقت مع التجلط الوريدي الجيبي المخي، ونستعرض في هذا المقال حالة مريض مصاب بمرض كرون وحدث لديه تجلط في الوريد الدماغي العميق، وتجلط سهمي جيبي. لقد شُخصت الحالة في قسم العناية المركزة بمستشفى الملك فيصل في مكة المكرمة، المملكة العربية السعودية.

Inflammatory bowel disease (IBD) is an idiopathic inflammatory disease of the gastrointestinal tract. There is a well-known risk of thrombosis in patients with IBD with an overall incidence of 1.3-7.5%. Cerebral venous sinus thrombosis (CVST) has been reported as an uncommon complication of IBD. It is suspected to be a consequence of genetic predisposition and the hypercoagulable state occurring during periods of increased activity. There are a few cases of Crohn's disease in the literature associated with CVST. Here, we describe a patient of Crohn's disease with thrombosis of the internal cerebral vein and sagittal sinus thrombosis in the intensive care unit of King Faisal Hospital, Makkah, Kingdom of Saudi Arabia.

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Inflammatory bowel disease (IBD) comprises ulcerative colitis (UC) and Crohn's disease (CD). Neurologic manifestations are particularly severe and include thrombotic and cerebrovascular disease, myelopathy, cerebral vasculitis, multiple sclerosis, and acute disseminated encephalomyelitis.[1] The incidence of cerebral venous sinus thrombosis due to inflammatory bowel disease ranges from 1.3-7.5%,[2] and is usually due to the hypercoagulable state that can occur in these patients during the relapse of the disease. The most frequently involved vessels are transverse sinus (86%), superior sagittal sinus (62%), straight sinus (18%), cortical veins (17%), vein of Galen (16%), and internal cerebral veins (11%).[3] Thrombosis of the great cerebral vein is a form of stroke due to a blood clot in the vein. It affects just 3-8% of patients, predominantly women.[4] Thrombosis of the cerebral veins and sinuses accounts for less than 1% of all strokes.[5] Eighty-five percent of the patients show either acquired or inherited prothrombotic risk factors. Inflammatory bowel diseases such as Crohn's disease and ulcerative colitis are described as risk factors for venous thrombosis.[6] Our aim in presenting this article is to highlight that CVST is an elusive diagnosis because of the nonspecific presentation and numerous predisposing factors. Imaging plays a key role in the diagnosis.

Case Report. A 48-year-old female patient, a known case of Crohn's disease on immunosuppression presented with a history of gradually worsening severe headache, vertigo, and slurring of speech of 2 days duration. On admission she had left sided hemiparesis and altered level of consciousness. She had one episode of vomiting and diarrhea on the day of headache. Her vital signs were as follows: temperature 38.5°C, pulse 96/minute, blood pressure 140/80 mm Hg, and respiration 22/minute. Laboratory investigations were as follows: white blood count 13,600/cmm (normal range: 4000-10,000/cmm), hemoglobin 13.8 g/dL, platelets 2.7 lacs/cmm, random blood sugar 96 mg/dl, urea 19 mg/dl, creatinine 0.8 mg/dl, sodium 138 mEq/L, potassium 3.6 mEq/L, aspartate aminotransferase 18 IU/L, alanine aminotransferase 13

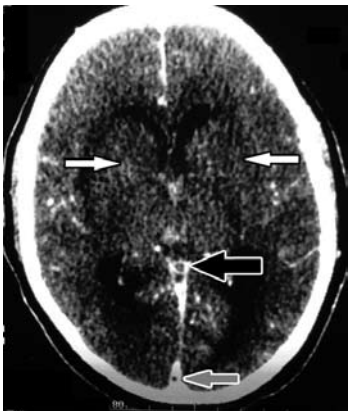

Figure 1 - A CT scan of the brain with contrast showing hypodense basal ganglia (white arrow), a filling defect in the great vein of Galen (black arrow), and another filling defect posteriorly (grey arrow) in the superior sagittal sinus giving the typical appearance of an empty delta sign.

IU/L, alkaline phosphatase 84 IU/L, coagulation profile was within normal range (prothrombin time 12 sec, partial thromboplastin time 28.5sec, INR 1.0).

Cardiovascular examination was as follows: S1, S2 present with normal sinus rhythm. Respiratory examination was as follows: bilateral equal air entry, and normal vesicular breath sounds. Her abdomen was soft, with no palpable lymph nodes or organomegaly. The CNS examination showed stupor, left sided hemiparesis, mildly brisk tendon reflexes on the left side, normal plantar reflexes bilaterally, pupils were bilaterally equal and reacting to light. Single slice CT brain imaging without contrast was carried out on admission (image not available), which showed bilateral hypodense basal ganglia and thalamus. Contrast study was carried out immediately, which showed hypodense basal ganglia, with filling defects within the internal cerebral veins extending to the great cerebral vein of Galen. Another filling defect was noted in the superior sagittal sinus posteriorly (grey arrow) giving the typical appearance of an "empty delta sign" suggestive of sagittal sinus thrombosis (**Figure 1**). Lumbar puncture was not carried out in view of cerebral edema. The CT findings concluded the diagnosis of cerebral venous sinus thrombosis with Crohn's disease, and due to lack of further interventional facilities she was referred to a higher center for further management.

Discussion. The etiology of hypercoagulation and thromboembolism in IBD remains poorly understood. Coagulation factor abnormalities such as elevated fibrinogen level, factor V, factor VIII, increase in circulating thrombin-antithrombin complexes, and decreased antithrombin III have been described; thrombocytosis and increased platelet aggregation have also been documented.[7] An alteration in the mucosal hemostasis, increased endothelial, and tissue factor activation are present in IBD. A CVST should be suspected in a patient who presents with recent onset of unusual increasing headache, seizures, or stroke, without any predisposing factors. Indirect CT signs include parenchymal abnormalities (pre contrast), focal enhancement (post contrast), and small ventricles compressed by the cerebral edema. Bilateral cerebral involvement and infarctions in the nonarterial distribution with areas of hemorrhage should lead to suspicion of venous thrombosis. The empty delta sign, which may be seen 5 days to 2 months from onset,[8] is the most frequent direct sign of CVST and can be seen on enhanced CT scan. It represents a filling defect in the dural sinus and is due to: (1) recanalization of the thrombus within the sinus, (2) organization of the clot, (3) blood-brain barrier breakdown, and (4) dilatation of collateral peridural and dural venous channels.[9] However, the sensitivity of the CT scan for such diagnosis remains 68%. An MRI in conjunction with MR venography are considered the best noninvasive tools for diagnosis and follow-up.[10] The 2 main factors contributing to the thrombosis in IBD are the presence of hypercoagulable state in inflammatory bowel disease,[11] and the corticosteroid induced hypercoagulable state.[12] The CT venography also can confirm the diagnosis. The differential diagnosis, which must be ruled out, includes acute stroke, subarachnoid hemorrhage, neurosarcoidosis, and systemic lupus erythematosus.

In conclusion, cerebral venous thrombosis is a known complication of inflammatory bowel disease and due to nonspecific clinical findings neuroimaging plays a major role for delineating the thrombus. The key to the diagnosis remains bilateral cerebral involvement, and infarction in the non- arterial distribution with areas of hemorrhage.

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