Cerebral Vein Thrombosis in a Child with Crohn's Disease

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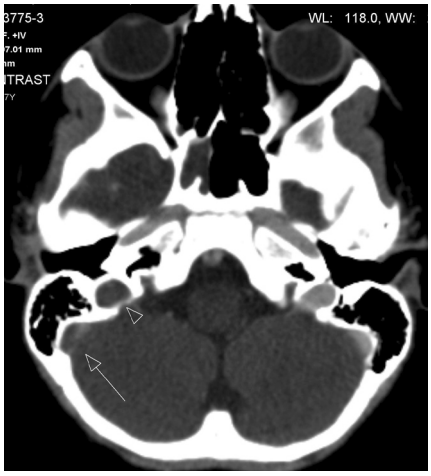
Inflammatory bowel disease is associated with an increased risk of vascular complications, the most important of which is thromboembolism. The propensity of patients with IBD to thromboembolic events is partly attributed to the existence of a hypercoagulable state, especially during periods of active disease [1]. Vascular complications of Crohn's disease are quite rare in children, and there are only a few reports of this association in the literature. We describe a 7 year old patient who was diagnosed with active Crohn's disease after he was diagnosed with thrombosis of various cerebral sinuses and pulmonary arteries.

Patient Description

A 7 year old boy was admitted for evaluation of microcytic anemia, headaches and fever. His main complaints were fatigue, frontal and right periorbital headaches with alternating blurred vision, and episodes of recurrent vomiting accompanied occasionally by watery diarrhea. There was no history of arthralgia, arthritis or skin involvement. Microcytic anemia, which had been noted 4 years before admission, did not respond to iron supplementation. The family history revealed that the patient's mother has an epileptic disorder, his father suffers from multiple sclerosis, a grandmother had died from intracranial bleeding when she was 24 years old, and an aunt had unspecified anemia.

On admission, the patient looked ill and pale and was febrile. Blood pressure was 95/50 mmHg, heart rate 100/min, and oxygen saturation 99%. Ophthalmological examination was normal and there were no abnormal neurological findings in the physical examination. Blood tests revealed microcytic anemia and low iron level, thrombocytosis and leukocytosis, as well as hypoalbuminemia (3.1 g/dl) and normal prothrombin time and partial thromboplastin time. C-reactive protein was 110 mg/L. Urine tests and a chest radiogram were normal.

Abdominal ultrasonography showed significant thickening of the walls of the terminal ileum and proximal colon, without hepatosplenomegaly or mesenteric lymphadenopathy. Because his headaches and eye pain persisted, a repeat ophthalmological examination was performed, revealing a bilateral nasal elevation of the optic nerve head. A contrast enhanced brain computed tomography scan demonstrated extensive sinus vein thrombosis which included the sagittal and right transverse and sigmoid sinus veins with extension to the right internal jugular [Figure]. A lumbar puncture showed an elevated intracerebral pressure (27 cmH_2O), but the cerebrospinal fluid was normal (no cells, 8.3 mg/dl protein, and 68 mg/dl glucose). Treatment with enoxaparin and acetazolamide was initiated. Further evaluation included CT scans of the chest and the abdomen to rule out neoplastic disorder. Chest CT scan showed thrombi in the pulmonary trunk, left and right main pulmonary arteries and in segmental arteries to both lower lobes. There were no enlarged lymph nodes or masses. Abdominal CT scan showed mural thickening of the terminal ileum with proximal ileal dilatation and mesenteric vessel congestion. There was blurring of the fat around the ascending colon and its wall was thickened. A few mesenteric lymph nodes were slightly enlarged in the right lower quadrant of the abdomen.



Contrast-enhanced CT of the head, demonstrating thrombus in the jugular vein (arrowhead) and in the sigmoid sinus (arrow)

As the patient's clinical condition stabilized, colonoscopy was performed. Colitis and terminal ileitis with pseudopolyps were observed. Treatment with intravenous corticosteroids was initiated, and after 24 hours the child was free of headaches and his body temperature returned to normal. The histopathological findings from the biopsies that were taken by colonoscopy confirmed the diagnosis of Crohn's disease. Thrombophilia screening essays showed that the patient was homozygous for the MTHFR mutation and heterozygous for the prothrombin mutation. Screening was normal for protein S, protein C activity, antithrombin III activity, lupus anticoagulant and factor V Leiden.

IBD = inflammatory bowel disease

Comment

Thromboembolism is a recognized disease-specific extra-intestinal manifestation of inflammatory bowel disease [2]. The most common events are deep vein thrombosis of the leg and pulmonary embolism [2]. It has been suggested that disease activity and the extent of colonic localization are correlated with the risk of developing thromboembolism [2,3]. Both fibrinolysis and coagulation are activated in IBD, especially during active disease. A recent study investigating the role of prothrombin gene mutations in the occurrence of thrombosis in IBD patients revealed that it is mostly acquired risk factors that play a role in thromboembolic events in IBD patients, particularly during active phases of the disease [3], while inherited risk factors are found less frequently. Reports of thromboembolism in pediatric IBD are rare, and most of them occurred in older children with ulcerative colitis [4]. In children, as in adults with IBD, a genetic predisposition to hypercoagulability does not appear to correlate with the occurrence of thromboembolic events. Our patient demonstrated both an acquired and genetic predisposition for thrombophilia. Although Crohn's disease was diagnosed immediately after the diagnosis of cerebral sinus vein thrombosis (on the same admission), the patient's intestinal disease had already been active for at least several months. The presence of a genetic hypercoagulability is in itself not considered a prominent risk factor for thrombosis in IBD patients in comparison to a non-IBD population with these mutations [5]. Nevertheless, our patient was most probably predisposed to thrombotic complications by the combination of his genetic susceptibility together with the active bowel disease. Pediatricians should be aware of these serious complications even in young children with Crohn's disease and central nervous system manifestations.

References

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