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Case Study

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VERY EARLY ONSET- INFLAMMATORY BOWEL DISEASE (VEO-IBD) IN A THREE YEARS OLD CHILD: A CASE REPORT

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BACKGROUND

Inflammatory bowel disease (IBD) is a chronic inflammatory condition of the gastrointestinal tract, and its pathogenesis involves genetic and environmental factors. Inflammatory bowel disease (IBD) that presents in children <6 years of age is known as very early-onset IBD (VEO-IBD). Since childhood-onset IBD seems to be a more aggressive and rapidly progressive disease than adult-onset IBD, it should therefore be diagnosed and treated immediately. This subgroup is often characterized by higher severity, aggressive progression, strong family history of IBD, predilection for colon-only involvement, and poorer response to conventional treatments. Immunodeficiencies are identified in up to 25% of cases, which may impact response, safety, and indication to different therapies. Here, we report a case of very early-onset IBD (VEO-IBD) in a three years old boy with clinical and biochemical manifestations. The diagnosis was confirmed with histopathological evidence.

INTRODUCTION

Very early-onset IBD (VEO-IBD) is a designation given to disease diagnosed before 6 years of age.[1,2] Approximately 6 to 15% of the pediatric IBD population presents at <6 years of age, including, although rare, children diagnosed in the first year of life.[3] The phenotype of VEO-IBD is heterogeneous and while some children have mild disease, others can present with more extensive and severe disease than older onset pediatric and adult IBD.[4-7] Due to the more aggressive phenotype, early age of onset, and strong family history, a subset of VEO-IBD is now considered to be a monogenic disease, often involving genes associated with primary immunodeficiencies.[8-10] and approximately 50 genetically different mutations have been identified using advanced genetic sequencing techniques.[9-10] IBD should be considered in the differential diagnosis of any child with persistent diarrhea, hematochezia, failure to thrive, and/or poor feeding. We report the case of a three-years-old Bangladeshi boy who presented with diarrhea and hematochezia and was finally diagnosed as VEO-IBD and treated. He showed good improvement with treatment.

CASE REPORT

Our patient Momen Hasan,3 years old boy, only issue of non-consanguineous parents admitted in BSMMU with the complaints of passage of loose stool for 2 years which was oily, 10-12 times/day, foul smelling, sometimes blood mixed .He is not growing well and lost weight during his course of illness. He had no past medical history and relevant family history. He is immunized as per EPI schedule. Also, the development was appropriate as per his age. He was formula-fed since birth. The boy was physically examined, and the results revealed that he was ill looking, irritable, pale, had no dysmorphic features, and had no dehydration. He is severely wasted and stunted (WAZ: -4.24; WLZ: -4.16). His abdominal examination had been normal except peri anal fistula (figure 1). The rest of the examination was unremarkable.

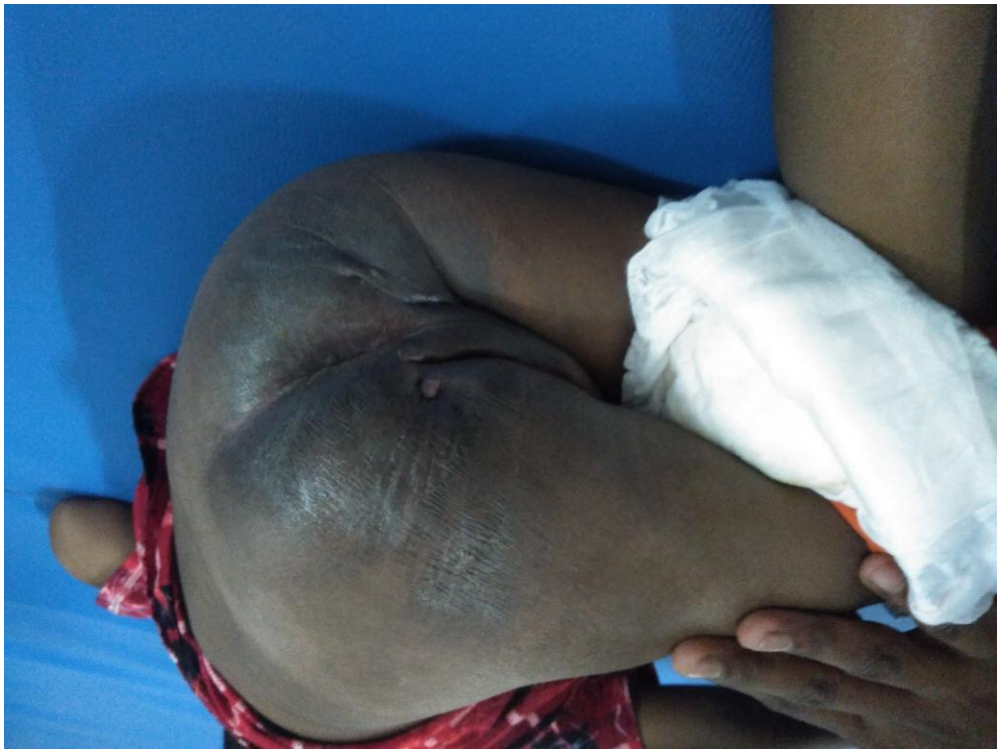

Figure 1: Showing multiple perianal fistula of Momen.

Laboratory results demonstrated, Hemoglobin: 8.2 gm/dl, Total WBC count 15,000/cmm, platelet count: 5,30,000/cmm, elevated inflammatory markers (ESR: 120 mm in 1st hour; CRP: 165 mg/L) and low albumin (16g/L), with a negative infectious work-up, including blood culture, stool culture, Clostridium difficile toxin, and viral serology (CMV). His Stool for fecal fat was negative and sweat chloride test was also negative for Cystic fibrosis. There was no improvement of bloody diarrhea on initial intravenous (IV) broad spectrum antibiotic coverage.

An abdominal ultrasound revealed edema in the descending and transverse colon. His immunological screening results were normal. We ruled out cytomegalovirus (CMV) infection, food protein-induced enterocolitis syndrome (FPIES), and primary immunodeficiency disorders based on the laboratory finding. On the fourteenth day of hospitalization, a colonoscopy revealed ulcers and pus in the rectum and descending colon associated with loss of mucous membrane from the rectum to the middle of the transverse colon. The histopathology report revealed crypt architectural abnormalities and apoptosis, moderate to severe chronic patchy active colitis, suggestive of IBD. Thus, the patient was diagnosed with VEO -IBD.

The patient's symptoms and laboratory findings improved in response to the treatment with mesalazine and prednisolone for one month; no relapse was observed after steroid taper. Azathioprine was added during the period of steroid tapering and maintenance treatment was continued with mesalazine and azathioprine. His clinical condition was under control with mesalazine and azathioprine. His physical development was appropriate for his age now.

DISCUSSION

Children who are diagnosed with IBD in the first 2 years of life are often referred to as infantile onset IBD, and those diagnosed between 2 and 6 years of age are classified as VEO-IBD. Approximately 40% of children with infantile and VEO-IBD have extensive colonic disease (pancolitis) at presentation.[4,7] however, the extent and location of disease can change and progress, making it difficult to differentiate ulcerative colitis (UC) from Crohn disease (CD). For example, initial isolated colonic disease can extend overtime to include the small bowel.[11] Furthermore, while the endoscopic findings often show a colonic distribution of disease, over time, the histology in some of these children can change and demonstrate features consistent with CD, such as granulomas or duodenal villous blunting. These findings can have important implications when determining the appropriate surgical approach in patients with severe colitis. Therefore, IBD-unclassified (IBD-U) is diagnosed more often in patients with VEO-IBD (11%–22%) as compared with older onset IBD (4%–10%).[12-16]

Treatment approaches for pediatric patients sometimes differ from those for adult patients. To date, all effective therapies in adults have also been effective in children. This is a great need for clinical trials of new therapies in children so that they have equal access to emerging treatments and optimal pediatric dosing can be established.[14]

Our patient experienced severe diarrhea and hematochezia; the differential diagnosis included infection, allergic colitis and lymphoid follicular hyperplasia, although all tests for these disorders were negative. Colonoscopy and pathological findings of intestine specimens confirmed the diagnosis of VEO-IBD, and treatment with mesalazine and steroids was effective at promoting resolution. We initially administered mesalazine 50 mg/kg/day and prednisolone 1 mg/kg/day. Long-term steroid use is not preferable for pediatric patients due to its side effects such as disturbance in growth, osteoporosis, and diabetes. Hence, steroid use is recommended only for a short period in children. When these treatments are deemed ineffective, immunosuppressive therapy or stem cell transplantation should be considered as an alternative.

In this case, genetic mutation study could not be done as genetic mutation analysis for IBD is not available in Bangladesh. Approximately 50 genetically different mutations have been identified with monogenic IBD using advanced genetic sequencing techniques. With further research, new biomarkers and advanced diagnostic techniques used in the field of gastrointestinal endoscopy, molecular pathology, and genetics need to be developed for establishing appropriate diagnosis and treatment.

CONCLUSIONS

In conclusion, VEO-IBD should be included in the differential diagnosis of pediatric patients presenting with persistent diarrhea and hematochezia, and colonoscopy should be performed as soon as possible when VEO-IBD is suspected in order to facilitate timely treatment.

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