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Case Report

Cystic fibrosis presenting as small bowel obstruction in pediatric patient: a case report

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ABSTRACT

Intestinal obstruction complete or incomplete has been found commonly in patients with cystic fibrosis in all age groups with variable presentation. Prevalence of intestinal obstruction in children with cystic fibrosis is 7-8%. Neonates present with meconium ileus due to inspissated meconium and complete obstruction. This case demonstrates similar presentation in a pediatric patient. A six day old male patient presented in the pediatric surgery department with abdominal distention, vomiting and fever. Patient was operated and intraoperative meconium plug was seen along with dilated proximal bowel, filled with sticky meconium. Resected gut segment was received in the Department of Pathology. Grossly dilated gut segment on cut section was seen filled with thick putty like greenish fecal matter. Histopathological examination showed features consistent with cystic fibrosis. There are multiple causes for intestinal obstruction in neonates, cystic fibrosis can be one of them which needs to be considered by the clinicians. Such cases need to be reported for better understanding of their clinico-pathological presentation and prevalence.

Keywords: Cystic fibrosis, Obstruction, Meconium ileus

INTRODUCTION

Cystic fibrosis is result of a defect in the cystic fibrosis transmembrane regulator (CFTR), which is responsible for the excretion of salt. The defects result in viscous secretions in multiple organs.[1] Pulmonary involvement by cystic fibrosis being the most common manifestation it has been historically considered to be a pulmonary disease.[2] With the increasing life expectancy gastrointestinal manifestations have become more important Gastrointestinal manifestations have a wide range of presentation due to mucus inspissation and dysmotility including GERD, MI, DIOS, intussusception and bacterial overgrowth.[3] Intestinal obstruction complete or incomplete has been found commonly in patients with cystic fibrosis (CF) in all age groups with variable presentation. Prevalence of intestinal obstruction in children with cystic fibrosis is 7-8%.[4] Neonates present with meconium ileus (MI) due to inspissated meconium and complete obstruction. This case demonstrates similar presentation in a pediatric patient.

CASE REPORT

A six day old male child presented in the pediatric surgery department with abdominal distention, vomiting and fever. Conservative management of his intestinal symptoms was not successful. Hence the patient was operated. Intra-operatively, dilated proximal bowel was seen with meconium plug in the distal segment. The gut segment was resected and received in the department of pathology in 10% formalin.

Findings

Grossly, a dilated segment of gut measuring 5 cm in length was received. External surface was unremarkable. On cut section distended lumen was seen filled with thick putty like greenish fecal matter. Representative sections were taken, processed and stained with H and E and PAS.

Microscopic examination revealed focal erosion of the lining epithelial cells with plugging of the mucosal crypts with mucin. Histopathological features were consistent with clinical diagnosis of cystic fibrosis and further molecular studies were advised for confirmation.

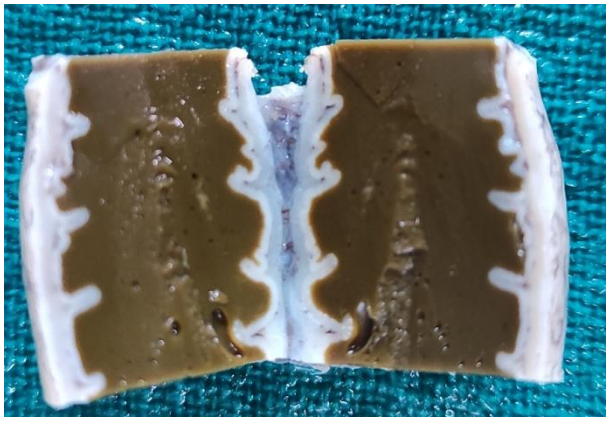

Figure 1: Gut segment filled with thick putty like greenish fecal matter.

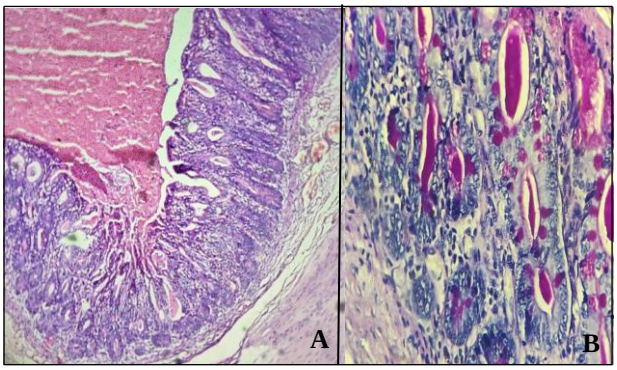

Figure 2: Erosion of the lining epithelial cells along with plugging of the mucosal crypts with mucin (A) H and E at 20X; and (B) PAS at 40X.

DISCUSSION

MI is associated with CF and leads to the intestinal obstruction due to thick meconium plug. In one of the study, of all the patients with CF, 13-17% present with meconium ileus, requiring surgical intervention in most of them.[4] Patients presenting with MI can be uncomplicated when presenting with accumulation of the inspissated meconium and complicated when intestinal obstruction is caused by meconium laden loops of bowel twisting and becoming ischemic leading to volvulus, perforation or atresia.[5] Diagnosing and confirming the histological appearance of CF can help to put the patient on proper treatment and avoid the associated complications. Treatment of meconium ileus is complicated. As seen in some studies the conservative treatment has 36-39% success rate and surgical management can result in higher complication rate than delayed anastomosis.[6] This patient was just 6 days old and the symptoms were not relieved by conservative management and hence was resorted to surgical intervention. Unfortunately the patient succumbed three days post-op.

CONCLUSION

There are multiple causes for intestinal obstruction in neonates, cystic fibrosis can be one of them which needs to be considered by the clinicians. Such cases need to be reported for better understanding of their clinico-pathological presentation and providing proper timely treatment.

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