A rare case of predominantly muscular infiltrative eosinophilic gastroenteritis with ascites: A case report and review of the literature

Introduction

Eosinophilic gastroenteritis, first reported by Kajiser in 1937, is a rare disorder that can present with various gastrointestinal manifestations, depending on the site of the affected gastrointestinal wall. Klein et al. have demonstrated that this disorder may be histopathologically classified into three major types: predominant mucosal, predominant muscle, and predominant subserosal layers. However, its clinical features, aetiology, and treatment have not yet been definitely established. We report a rare case of eosinophilic gastroenteritis with features of the predominant muscle layer type, with ascites, abdominal distension, diarrhoea and high grade fever.

Case Report

A 45-year-old patient presented with complaints of abdominal distension, occasional vomiting, high grade intermittent fever and increased frequency of stool for one month. There was no significant history of any other illness. He had a normal appetite and no allergy to any food item. There were no positive findings on general examination. On per abdominal examination, the abdomen was distended, soft, and non-tender, with no guarding or rigidity; the liver and spleen were not palpable, suggesting the presence of intra-abdominal free fluid. On per rectal examination fullness was appreciated. His complete blood was evaluated in which total counts rose to 15291/µL, absolute eosinophilic counts were 8410/µL, 55% of the total WBC count. His urine analysis, renal functions and liver functions were normal. The chest x-ray revealed right pleural effusion and x-ray abdomen in upright position showed ground glass appearance with few air fluid bowel loops. The ultrasound abdomen and thorax suggested moderate ascites and thickened bowel loops. CT scan confirmed the ultrasound findings and additionally showed diffuse thickened mucosal folds in the entire small bowel loops. Diagnostic ascitic tap was carried out which reported reddish yellow turbid fluid; the cell count was 8060 cells/µL in which the polymorphs were 90% and lymphocytes 10%. Smears with gram and ZN stains were negative. Ascitic fluid culture was negative. Ascitic fluid adenosine deaminase was negative. On cytological examination no malignant cells were seen. He underwent upper gastrointestinal endoscopy and sigmoidoscopy to rule out infiltrative disease and storage disease which were not conclusive on histopathology. Then it was decided to take full thickness biopsy from the bowel, peritoneum and from the liver through diagnostic laparoscopy. On laparoscopy there was moderate ascites with diffusely dilated small bowel. Biopsies were taken from liver, peritoneum and jejunum. On histopathological examination liver tissue was normal, peritoneal biopsy showed infiltration of the tissue by chronic inflammatory cells along with eosinophils; jejunal biopsy consisted chiefly of the muscularis propria which was infiltrated by eosinophils. Thus his diagnosis confirmed eosinophilic gastroenteritis. He was prescribed prednisolone 20 mg once a day for 6 weeks which was then tapered off. His ascites was relieved within 15 days, clinical improvement noted within one month and he was followed up for one year after stopping the drug and did not show recurrence of symptoms.

Discussion

Eosinophilic gastroenteritis was first reported by Kajiser in 1937. In 1970, Klein1 classified the disease according to the predominance of eosinophilic infiltration in the different layers of the intestinal wall. Talley et al.2 identified four main diagnostic criteria: (1) the presence of gastrointestinal symptoms, (2) biopsies demonstrating eosinophilic infiltration in one or more parts of the gastrointestinal tract, (3) absence of eosinophilic involvement of multiple organs outside the GI tract and (4) no evidence of parasitic or extra-intestinal disease. Disease is rare, undiagnosed and surely underreported.2,3 Patients typically present in the third through fifth decades of life, but the disease can affect any age group. An equal gender distribution or a slight male preponderance has been reported.3 The cause is unknown and pathogenesis is poorly understood. Any segment of the GI tract may be involved, including the oesophagus4–6 or colon,6 but
most commonly the stomach or small bowel is affected. Classification is based on the layer of the gut wall primarily affected. Most prevalent form is characterised by mucosal (and submucosal) disease.1,23 Predominant mucosal layer disease presents with pain, nausea, vomiting, diarrhoea, weight loss, iron deficiency anaemia, malabsorption, and protein loosing enteropathy, whilst predominantly muscle layer disease presents with the obstructive features. The rarest form is serosal disease; all layers of the bowel are usually involved, and patients present with typical ascites.1,2,3 Only rarely is mucosal involvement absent where serosal disease is present. However, the definitive diagnosis of eosinophilic gastroenteritis requires histological evidence of eosinophilic infiltration. Blood investigation shows peripheral eosinophilia in about 80% cases; the absolute eosinophil count averages 1500–2000 cells/µL in patients with disease of the muscle layer, although the count may fluctuate markedly over time.3 Other findings include iron deficiency anaemia, hypoalbuminemia. Radiological changes found in eosinophilic gastroenteritis are variable, nonspecific, and absent in at least 40% of patients.8,9 The gastric folds are enlarged, with or without nodular filling defects. CT may demonstrate thickened intestinal wall and localised mesenteric lymphadenopathy; with serosal involvement; ascitic fluid is usually detected. When subserosal disease involves the small bowel, biopsy of the mucosal layer taken during gastroscopy or colonoscopy often fails to diagnose eosinophilic gastroenteritis. Laparotomy or laparoscopic full thickness biopsy of the bowel is often required for confirmation in such cases. On histopathology it is characterised by oedema and an inflammatory cell infiltrate that is almost entirely composed of eosinophils; the eosinophils may occur in clumps.10 Treatment with steroids is the mainstay in the management of eosinophilic gastroenteritis. Other modalities include diet modification, anti-helminthic, and mast cell stabilisers (sodium cromoglycate). Surgical intervention is required only when a definitive diagnosis cannot be made or when complications like obstruction, perforation or bleeding occur. Because the natural history of eosinophilic gastroenteritis has not been well documented, long-term follow-up is required.

References