

# Primary Eosinophilic Gastrointestinal Disorders: An Update of Presentations

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## Abstract

Primary eosinophilic gastrointestinal disorders (EGID) affect mainly the stomach and duodenum. Other organs in gastrointestinal tract are rarely affected. It is a rare clinical entity with no definitive etiology. The varied presentations of EGID account for anxiety of surgeons. The most common presentation of EGID is pain in the abdomen, and high index of suspicion is required for the proper diagnosis of this entity in view of treatment differs on its presentations. In this short communication, we highlight the rare and unusual presentations of EGID.

**Key words:** Eosinophilia, Gastroenteritis, Gastrointestinal diseases

## INTRODUCTION

Eosinophilic gastrointestinal disorders (EGID) referred to eosinophilic gastroenteritis (EG) which is a group of disorders including eosinophilic esophagitis, gastritis, enteritis, and colitis. EGID is a term coined by Rothenberg<sup>1</sup> in 2004.

EGID is an uncommon gastrointestinal tract (GIT) disease and challenging task to diagnose. Physicians must maintain a high index of suspicion and a working knowledge of the natural history of EG is important to establish the proper diagnosis.<sup>2</sup> The symptoms and signs of EGID can simulate an acute abdomen, acute appendicitis, obstructive cecal mass, pancreatitis, cholecystitis, duodenal ulcer, and intussusception.<sup>2</sup> The combination of acute/recurrent abdominal pain with peripheral and tissue eosinophilia confirms the diagnosis of EGID.<sup>2</sup> The etiology of EGID is unknown but the speculation has focused on the selective release of eosinophilic major proteins and causes intestinal epithelial damage.<sup>2</sup>

Over a period of 10-year of histopathology practice, we encountered the three unusual presentations of the primary

EGID. Primary EGID is defined as the involvements of GIT primarily with eosinophils rich inflammation in the absence of known cause of eosinophilia including drug reaction, parasitic infestations, and malignancy.<sup>3</sup> The most common form of EGID presenting with chief complaints of “mucosal” form as nausea, vomiting, diarrhea, acute abdomen, malabsorption, anemia, protein-losing enteropathy, etc. The “muscular” form presented with fibrosis due to thickening of bowel wall in view of diffuse infiltration of eosinophils in the muscular layer. We have two cases of this form in the small intestine and appendix, respectively.<sup>4,5</sup> Exudative ascites is the main presentation in the serosal form. As per Talley *et al.*,<sup>6</sup> peripheral eosinophilia has been reported in 80% cases of EGID. However, definitive diagnosis requires histopathological evidence of eosinophilic infiltrations.<sup>4</sup>

As per experience, first, we encountered with 54-year-old male presented to emergency department with acute abdominal pain and vomiting with no other positive and relevant history.<sup>4</sup> Abdominal X-ray showed multiple air-filled levels in the small intestine. Emergency laparotomy revealed multiple strictures at the ileum with proximal dilatation. Histopathologically, EGID was confirmed and strictures due to muscular form of EG.<sup>4</sup> Post-operative stool and occult blood test was negative.

The second case was 25-year-old male presented with the acute right iliac fossa pain below umbilicus.<sup>5</sup> Provisionally diagnosed and operated for an appendectomy. To our surprise, obliterative eosinophilic appendicitis was noted on

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histopathology. Primary obliterative appendicitis was even rare and presented with unexplained symptoms.<sup>5</sup>

The third case was even interesting with unusual presentation of EGID, eosinophilic appendicitis presented as mucocele.<sup>7</sup> Mucocele is a rare condition and often diagnosed as abdominal malignancy. The proper diagnosis of mucocele is only made on laparotomy and histopathology. To the best of our knowledge, this was the first case of acute eosinophilic appendicitis presented as mucocele.<sup>7</sup> The main precursor for the mucocele formation is the obstruction of the lumen due to muscular thickening and fibrosis.<sup>7</sup>

Steroids, Na cromoglycate, ketotifen, and montelukast are the mainstay of medical line of treatment. Complicated cases like ours with obstruction and perforation requires surgical intervention with resection and end to end anastomosis.<sup>4</sup>

## CONCLUSION

EGID are a diagnostic dilemma due to varied clinical presentations. Most of the time investigations are only

contributory. The surgeons should be alert and vigilant with the unusual and rare presentations of EGID like stricture, obstruction, mucocele, and inflammation to avoid unnecessary surgical intervention.

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