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Case Report:

Primary Eosinophilic Obliterative Appendicitis

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Abstract: Primary eosinophilic appendicitis is a rare condition of unknown etiology having vague and unexplained symptoms. Histopathological hallmark of this entity is eosinophilic infiltration of the muscularis propria with accompanying edema separating the muscle fibers, and absence of neutrophilic infiltration. Preoperative correct diagnosis of this entity is not possible in view of lack of specific imaging technology. Histopathological examination is the gold standard for the diagnosis. Here, we present an unusual case of Eosinophilic Obliterative appendicitis in a 25 years old male patient.

Key Words: Eosinophilic appendicitis; Obstruction; Appendix; Eosinophils

Introduction:

Eosinophilic enteritis (EE) is an extremely rare disease, involving the entire gastrointestinal tract (GIT), although stomach and duodenum are frequently involved sites.[1] The pathogenesis and etiology of eosinophilic enteritis remains unclear.[1] Appendix is rarely involved by this entity.

Acute appendicitis is one of the most common surgical disease worldwide. It is primarily diagnosed on the basis of history and the physical examination with additional assistance from laboratory and radiographic findings.[2] Currently the most accepted explanation for the development of acute appendicitis is obstruction and secondary infection.[3] The obstruction is thought to be due to extra luminal adhesions or luminal causes such as faecoliths or lymphoid hyperplasia in the walls[3] or rarely as a result of the infective process. Primary obliterative appendicitis due to eosinophils in the absence of faecoliths or lymphoid hyperplasia is a rare clinical entity.

Herein, we report a case of primary eosinophilic obliterative appendicitis, which is an extremely uncommon clinical entity.

Case Report:

A twenty five years old male presented with chief complaints of recurrent pain in right iliac fossa just below the umbilicus. It was not associated with fever, vomiting or diarrhoea. There was no family history of allergy or atopy and no history of drug allergy, asthma or allergic rhinitis. Local examination revealed mild to moderate tenderness in right iliac fossa. Rest of systemic examination was not contributory. Routine hematological examination showed hemoglobin 12gm%, total leucocyte count 13100/cmm and differential count of neutrophils 80%, lymphocytes 13%, and eosinophis 3%. Other biochemical and serological examination were unremarkable.

The patient was clinically diagnosed as appendicitis and open appendicectomy was performed under general anesthesia and resected specimen was sent for histopathological examination. Post-operative period was uneventful and patient was fine on regular follow up.

Gross features: The appendicectomy specimen measured 5.5cm in length. External surface showed congested blood vessels and covered with grey-brown areas of thick exudates. On cut section showed obliteration of the lumen and thickened wall was seen. The obliterated lumen show grey whitish appearance.

Light microscopy: Multiple section studied shows appendix. Mucosa showed extensive areas of ulcerative and denuded epithelium (Fig. 1). Submucosa showed necrotic areas, congestion and oedema. All the layers including muscular propria showed dense and diffuse infiltration by eosinophils that was >25-50/hpf (Fig. 2). All the layers showed congestion and serosa showed fibrosed walls with dense, diffuse eosinophilic infiltration. Final histopathological diagnosis was given as Primary eosinophilic obliterative appendicitis.

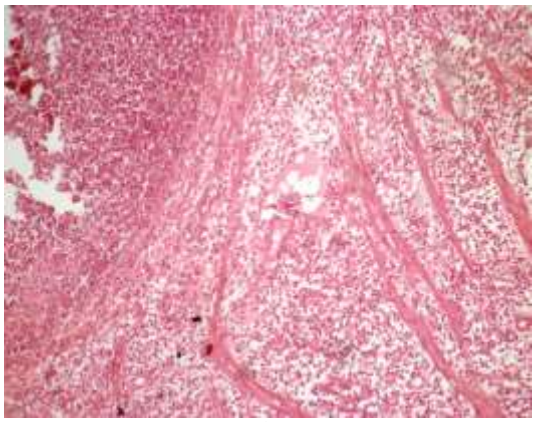

Figure 1: Photomicrograph of appendicular mucosa showed extensive ulceration of the epithelium with mild dense, diffuse infiltration of eosinophils in all coats (H & E, x100).

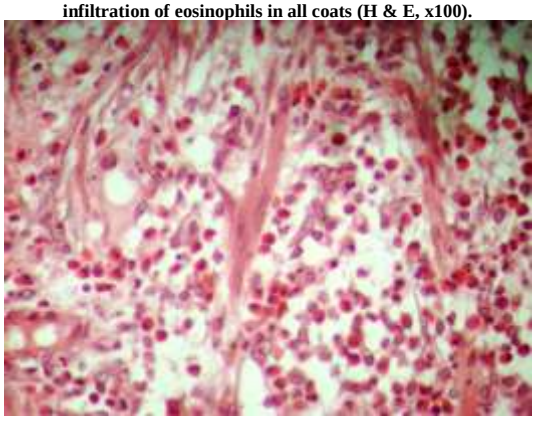

Figure 2: Photomicrograph showing muscularispropria with dense and diffuse infiltration by eosinophils (H & E, x400).

Discussion:

Primary eosinophilic appendicitis (EA) primarily affects the appendix with eosinophil rich transmural inflammation in excess of 10 per cmm (more than 25 eosinophils per high power field) in muscularis mucosa with absence of polymorphs or any other pathology in the wall as per Carr NJ[4], with no known cause for eosinophilia including drug reaction, parasitic infections and malignancy.[5]

Primary eosinophilic obliterative appendicitis is a rare clinical entity. The cause for obstruction of the appendicular lumen may vary as inflammatory stricture, appendicolith, villous adenoma, carcinoid tumour, mucosal web, endometriosis, carcinoma and extrinsic compression or inflammatory process. Usually a specific cause of obstruction may not be found as documented by Rokitansky CF.[6]Eosinophilic appendicitis may present as obstruction or rarely as mucocele[7] or eosinophilic edema.[8]

Depending upon the involvement of different layers of intestinal wall, symptoms may vary. The mucosal form of eosinophilic appendicitis, the most common variant, is characterized by vomiting, abdominal pain, diarrhoea, blood loss in stools, iron deficiency anemia and malabsorption. The muscularis form is characterized by infiltration of eosinophils predominantly in the muscle layer leading to thickening of walls due to fibrosis, resulting in obliterative appendicitis. This may have happened in our case. Serosal form is characterized by exudative ascites.[9] Carr NJ[4] suggests that an eosinophil count in excess of 10 per cmm (25 per 10 HPF) could be abnormal and labelled as eosinophilic appendicitis. Our case fulfilled these criteria. In our case, eosinophils were present in all the layers including muscularis propria (25-50 per HPF) leading to obliteration of lumen due to muscular fibrosis.

The pathogenesis and etiology of the disease is not well understood, hence no standard for the diagnosis of eosinophilic appendicitis exists. Tally et al[9] have identified three main diagnostic criteria for EE as:

1. Presence of gastrointestinal symptoms
2. Biopsies demonstrating eosinophilic infiltration of one or more sites of GIT.
3. No evidence of parasitic or extrinsic disease.

In our case, all the criteria were fulfilled. Peripheral eosinophilia was not noted in our case. Stool and upper and lower gastrointestinal scopy in our case was not significant. As per Tally et al[9], in 80% of the cases peripheral eosinophilia was noted. However gold standard of EE and EA requires histopathological evidence of eosinophilic infiltration.[7,9]

Mainstay of treatment is surgical intervention when complication as obstruction was evident. Steroids, sodium chromoglycate, ketotifen, montelukast may be medical alteratives.

Conclusion:

Primary eosinophilic obliterative appendicitis is a rare clinical entity with unexplained symptoms. Surgeons should think of this condition in differential diagnosis of abdominal pain. Histopathology is the gold standard for diagnosis of this rare condition.

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