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Case Report

Eosinophilic Appendicitis Presented As Mucocele: A Rare Clinical Entity

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ABSTRACT

Eosinophilic appendicitis (EA) is a rare disease of unknown etiology. Histological hallmark of this entity is eosinophilic infiltration of the muscularis propria with accompanying edema separating muscle fibers. Mucocele of the appendix is a rare condition that frequently mimics abdominal tumors. There is no specific imaging technology to correctly diagnose this condition preoperatively. So proper diagnosis of mucocele is only made during a laprotomy and histopathology. We herein present a case of eosinophilic appendicitis presented with mucocele, which is extremely rare finding. To the best of our knowledge, this is the first case in the literature of EA presented as mucocele.

Key words: Eosinophilic appendicitis, mucocele, appendix.

INTRODUCTION

Eosinophilic appendicitis (EA) is a rare clinical entity. It is characterized by acute presentation and grossly inflamed appendix with absence of neutrophils in muscle layer. Histological hallmark of this entity is eosinophilic infiltration of the muscularis propria with accompanying edema separating muscle fibers. [1] The gastrointestinal symptoms are dependent upon the predominant layer involved. [2] Diagnosis of eosinophilic appendicitis requires high index of suspicion and exclusion with various disorders that are associated with peripheral and tissue eosinophilia. [2] Mucocele is a descriptive clinical term for an abnormal mucous accumulation distending the appendicular lumen. [3] Mucocele is not diagnosed on

preoperative by imaging technology as many differential diagnoses. Hence it is a laparotomy diagnosis. [4]
We herein report a rare case of eosinophilic appendicitis presented with mucocele which is extremely uncommon.

CASE HISTORY

A thirty five year male came to surgical OPD with complaint of pain in right iliac fossa since two days. The pain was gradually increasing in intensity. It was not associated with fever, vomiting or diarrhea. There was no significant past, present or family history. Local examination revealed tenderness in right iliac fossa. Systemic examinations were found to be within normal limits. Routine hemogram showed hemoglobin 15 gm%, total leukocyte count 12,300/cumm and in differential count neutrophils 72%, lymphocytes 20% and eosinophills 08%.Rest biochemical and serological investigations were within normal limits. He had no history of drug allergy, asthma or allergic rhinitis. There was no history of allergy or atopy to family members. It was diagnosed clinically as acute appendicitis and open appendicectomy was done under general anesthesia and resected specimen was sent for histopathology. Post operative period was uneventful and patient is on regular follow up.

Gross examination:

Received an appendicectomy specimen measuring 7 cm in length. Externally appendix appeared swollen and dilated with congested veins and measured 3.5 cm in maximum diameter. (Figure 1) On cut sections showed dilatation of lumen filled with mucoid material with thickened wall. (Figure 2)

Fig. 1: Gross photograph of swollen and dilated appendectomy specimen.

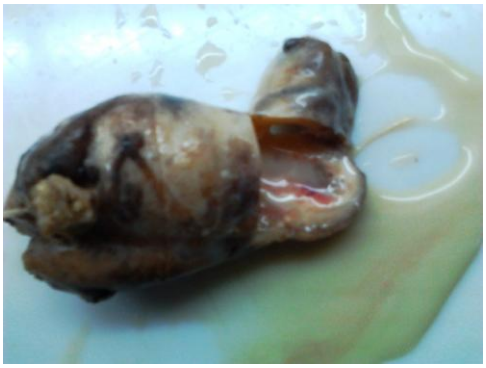

Fig. 2: Gross photograph of cut section of appendix showing dilatation of lumen filled with mucoid material with thick wall.

Light microscopy:

Multiple sections studied show appendix. Mucosa showed ulceration and flattening of epithelium.(Figure 3) Submucosa showed lymphoid follicle predominance. All the coats especially muscularis propria showed dense and diffuse infiltration of eosinophils (> 25-60%). (Figure 4) All of the coats show congestion and serosa showed fibrosed wall with dense diffuse eosinophilic infiltration. Final histopathological diagnosis was given as eosinophilic appendicitis with mucocele and eosinophilic peritonitis.

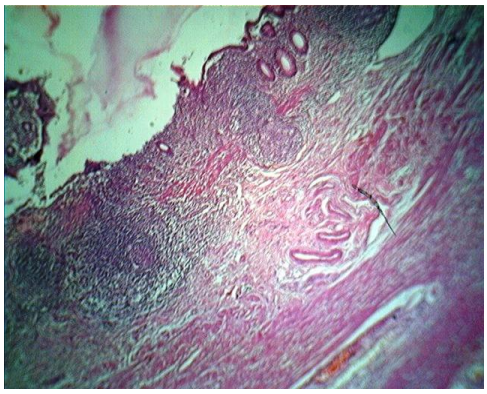

Fig. 3: Photomicrograph of appendicular mucosa showed ulceration and flattening of epithelium with mild dense, diffuse infiltration of eosinophils in all coats.(H & E, x100).

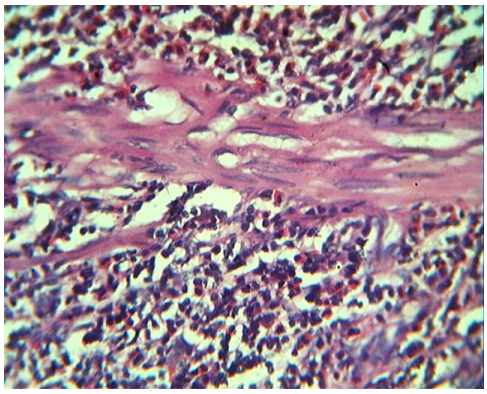

Fig. 4: Photomicrograph of muscularis propria showed dense and diffuse infiltration of eosinophils.(H & E, x400).

Post operatively we advised follow up CBC with absolute eosinophil count, upper gastro endoscopy to rule out stomach involvement and stool examination to rule out any parasitic infestations. During the follow up period of one year, upper GI endoscopy and biopsy was performed which showed no eosinophilic infiltration in the stomach. Post operative stool examination was negative for ova, cyst or worm on two separate occasions. Hematological examination was within normal limit after 2 to 6 weeks of steroid treatment.

DISCUSSION

Mucocele of the appendix is a nonspecific, descriptive clinical term for an abnormal mucous accumulation distending the appendicular lumen, regardless of the underlying cause. [3,5,6] It was first described by Rokitansky in 1842. [7] In 1940 Woodruff R and McDonald JR [8] classified appendiceal mucocele into benign type representing mucocele caused by obstruction of appendicular lumen and a malignant type representing mucin secreting adenocarcinoma. Clinical spectrum of all mucocele as mucinous neoplasm comprises mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma. [3]

Mucocele is a rare clinical disease. These lesions are found during appendectomy and constitute about 0.2 to 0.3 % of all appendectomies. [6] Eosinophilic appendicitis is extremely rare cause of mucocele.

The precursor for the formation of mucocele is obstruction of the appendicular lumen. [3] The causes for obstruction of the appendicular lumen may varied as inflammatory stricture, carcinoid tumor, villous adenoma, appendicolith, mucosal web, endometriosis, carcinoma, and extrinsic compression. Usually a specific cause of the obstruction may not be found as documented by Rokitanasky CF. [3]

Depending upon the involvement of different layers of intestinal wall, symptoms may vary. The mucosal form of eosinophilic enteritis (most common variant) is characterized by vomiting, abdominal pain, and diarrhea, blood loss in stools, iron deficiency anemia, malabsorption and protein loosing enteropathy. The muscularis form is characterized by infiltration of eosinophils predominantly in muscle layer

leading to thickness of wall due to fibrosis which may result in obstructive symptoms such as mucocele in our case of eosinophilic appendicitis. Serosal form is characterized by exudative ascitis. [2]

In our case, eosinophils were present in all the layers including muscularis propria leading to obstruction of lumen due to muscular thickness and fibrosis. Due to fibrosis, lumen was filled with mucinous material leading to mucocele as a late sequel. In our case, other causes of tissue and peripheral eosinophilia have been ruled out by history, upper GI endoscopy and stool examination.

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

i. Presence of gastrointestinal symptoms.
ii. Biopsies demonstrating eosinophilic infiltration of one or more areas of gastrointestinal tract.
iii. No evidence of parasitic/extrinsic disease.

In this case, all three criteria were satisfied. Peripheral Eosinophilia has been reported in upto 80% of the cases by Tally et al. [9] However, the definite diagnosis of eosinophilic enteritis requires histological evidence of eosinophilic infiltration. [2]

Treatment with steroid is mainstay of the treatment of eosinophilic enteritis and sodium chromoglycate, catotifen, montelucast may be tried. Complicated case with obstruction as in our case of mucocele or perforation requires surgical intervention. Otherwise surgeon should avoid unnecessary surgical intervention.

CONCLUSION

Eosinophilic appendicitis is a rare condition of unknown etiology with unexplained and vague symptoms. Hence surgeons should think of this condition in differential diagnosis of abdominal pain. It may rarely present as mucocele due to obstructive symptoms as in our case. To the best of our knowledge, this is the first case of eosinophilic appendicitis presented as mucocele in the literature.

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