



Acute Eosinophilic Appendicitis: Case report of three cases with brief review of literature

Rajeshwari K^{*1}, N.V. Dravid¹, Karibasappa GN², Akshay Surana¹

¹Department of Pathology ACPM Medical College, Dhule, Maharashtra, India

²Department of Public Health Dentistry, ACPM Dental College and Hospital, Dhule, Maharashtra, India.

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Abstract

Acute eosinophilic appendicitis (AEA) is a rare variant of appendix inflammation. The histologic hallmark of this entity is eosinophilic infiltration of the muscularis layer with accompanying oedema separating the muscle fibres with out neutrophilic infiltration.

To the best of our knowledge there are very few cases of eosinophilic appendicitis (EA) in the absence of any other abnormality reported in the literature. Hence, we made an attempt to study the cases of AEA and to draw the relevant conclusion about the disease pathogenesis. Out of total 159 appendectomy cases, three cases were found to have eosinophilic appendicitis (EA) and these cases were studied for clinical and pathological findings. Incidence of 0.02% (3/15) with male preponderance was found. We herein, present the cases of AEA which is rare variant and less understood entity.

***Corresponding author:**

Dr. Rajeshwari K, Assistant Professor, Department of Pathology ACPM Medical College, Dhule, Maharashtra, India
Email:ujwalgk@gmail.com; Mob : 91750768899

Introduction

Acute eosinophilic appendicitis (AEA) is a rare clinical entity. It is characterized by acute presentation and grossly inflamed appendix with absence of neutrophils in the muscle layer. The histologic hallmark of AEA is eosinophilic infiltration of the muscularis propria with accompanying oedema separating the muscle fibres.^[1] Eosinophilic appendicitis is less well understood disease entity which needs detailed study of reported cases and clear cut defining criteria.

There are very few studies with respect to observation of eosinophilic infiltration of the muscularis propria in the absence of any other abnormality.^[2,3] Hence in the present study an attempt was done to study the cases of AEA

Case Report

A total of 159 appendectomy specimens were received in the department of pathology during the study period of June 2013- January 2015. All the cases of appendicitis were screened and those cases fulfilling the criteria of eosinophilic appendicitis were included in the study. Criteria to diagnose EA used were transmural eosinophilic infiltrate in the wall of appendix, more than 25 eosinophils per high power field in muscularis mucosa, absence of polymorphs or any other pathology in the wall.^[4]

Routine investigation like complete blood count (CBC), peripheral blood smear, urine and stool examination was carried to rule out peripheral blood eosinophilia and parasitic infestation.^[5] Out of total 159 appendectomy specimens. Three cases of AEA were found accounting for an incidence of 0.02% (3/159). All the three patients were extensively studied by taking detailed clinical history, physical examination and relevant investigations to rule out any allergic pathology and worm infestation.

Case 1: Twenty five years female complained of recurrent pain abdomen in the right iliac region just below the umbilicus since 6 weeks. Clinically the case was diagnosed as sub-acute appendicitis. Grossly the appendix was enlarged, oedematous and congested. Cut section revealed patent lumen. On microscopic examination the case was categorized as AEA.

Case 2: Fourty year's male presented to surgical outpatient department with recurrent pain abdomen and vomiting since two weeks. Patient was evaluated for intestinal obstruction. Radiological examination showed signs of small bowel obstruction. Laprotomy was done with resection and anastomosis and also found that the appendix was inflamed, congested and edematous insitu, hence appendectomy was done.

Grossly the resected appendix was enlarged, congested and on cut section showed lumen obliteration. On histopathology the case fulfilled the criteria of AEA.

Case 3: Sixty seven years male patient presented with acute abdominal pain, vomiting and abdominal distention since two days. On admission to the casualty the patient condition started deteriorating and was haemodynamically unstable. Radiological evaluation showed features of pneumatosis intestinalis with haemoperitonium. Clinical diagnosis of intestinal perforation with signs of peritonitis was made. Emergency exploratory laprotomy was performed and revealed enlarged, oedematous and perforated appendix with haemoperitonium.

Grossly the resected appendix was oedematous, congested and enlarged measuring 5cm in length with perforation in the wall of the appendix measuring 0.1x0.2 cm. Cut section showed obliteration with mucous secretion (figure 1). On histopathology showed features of AEA (figure 2 and 3) with signs of peritonitis.

Post operatively all the three patients were advised follow up CBC with absolute eosinophil count, upper gastro- endoscopy of the gastrointestinal tract (GIT) for tissue eosinophilic infiltration and stool examination to rule out 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.

Age range of the patients was between 25-67 years (table 1). Mean age of presentation was 44 years. None of the case had history of allergic disorder or peripheral smear eosinophilia/tissue eosinophilic infiltration. In all the three cases stool examination for parasite was negative.



Figure 1: Gross photograph of appendectomy specimen showing enlarged, oedematous and obliterated lumen with mucous secretions.

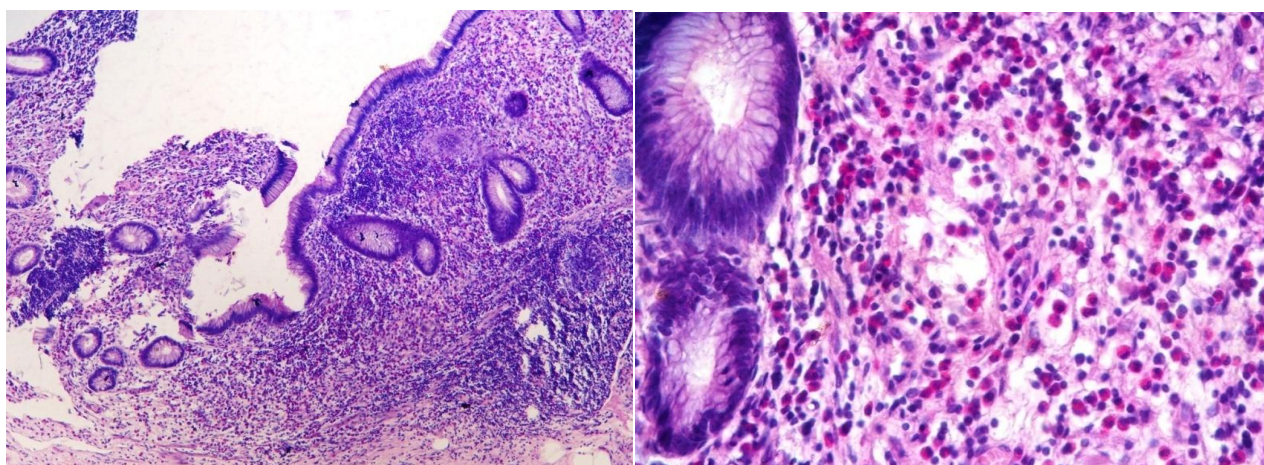


Figure 2 – Microphotograph of appendicular mucosa showing ulceration with dense and diffuse infiltration of eosinophils in all coats (H and E, x100).

Figure 3 - Microphotograph of muscularis propria showing dense and diffuse infiltration of eosinophils (H and E, x400).

Table 1 Clinical summary of three cases in acute eosinophilic appendicitis.

Case	Age/Sex	Clinical diagnosis	Peripheral blood eosinophilia	Stool Examination	Gross features
1.	25/female	Sub-acute intestinal obstruction	Absent	Negative	Oedema and congestion
2.	40/male	Intestinal obstruction	Absent	Negative	Oedema and congestion
3	67/male	Intestinal perforation with signs of peritonitis	Absent	Negative	Oedema congestion and perforation in the wall of appendix

Discussion

AEA was first proposed by aravindan in 1997 ^[6] and defined by aravindan et al in 2010. ^[1] He proposed that eosinophilic infiltration is an early event of appendicitis and represents the part of a type 1 hypersensitivity reaction to an allergen and primary pathologic changes characterized by eosinophilic oedematous lesion in the appendix. ^[1] These observations are novel and hypothesis requires further testing.

If the lesion is infected by bacteria, acute suppurative appendicitis occurs and if there is no infection, AEA occurs. The blood eosinophil count first increases and then decreases over time in cases with acute suppurative and eosinophilic appendicitis. ^[1] On the contrary, eosinophilia persists and does not resolve over time in eosinophilic gastroenteritis cases. ^[5] Thus, AEA should be evaluated as a variant of acute appendicitis rather than an extension of eosinophilic ga-

stroenteritis. ^[1, 6] Norman J Carr suggests that an eosinophil count in excess of 10 per mm² (25 per 10 HPF) could be abnormal. He also states differential diagnosis for this eosinophilic infiltrate as eosinophilic enteritis and infestation by parasites. ^[3]

In eosinophilic enteritis 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 losing enteropathy. The muscularis form is characterized by infiltration of eosinophils predominantly in muscle layer leading to intermittent obstructive symptoms and with complications like aspiration and perforation as in one of our case of eosinophilic appendicitis. Serosal form is characterized by exudative ascitis with intense peripheral eosinophilia. ^[7]

Because the pathogenesis and etiology of the disease is not well understood, no standard for the diagnosis of eosinophilic enteritis exists. Tally et al ^[5] have identified three main diagnostic criteria:

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

Peripheral eosinophilia has been reported in upto 80% of the cases by Tally et al. ^[5] However, the definite diagnosis of eosinophilic enteritis requires histological evidence of eosinophilic infiltration. ^[7] Steroid is the mainstay of treatment of eosinophilic enteritis and sodium chromoglycate, catotifen, montelukast may be tried. Complicated case with obstruction and perforation requires surgical intervention. Otherwise surgeon should avoid unnecessary surgical intervention in case of eosinophilic enteritis. In case of parasitic infestation it results in tissue injury and local irritation in the gastrointestinal tract and cure is possible with medical therapy. Hence once eosinophilic appendicitis is being diagnosed, the patient should be completely evaluated for eosinophilic enteritis and parasitic infestations.

In our study all the three cases showed eosinophils in all the layers including muscularis propria leading to obstruction and perforation. None of the case revealed peripheral blood eosinophilia/tissue eosinophilic infiltration of >1 site of GIT which was ruled out by history, peripheral blood smear examination and by upper GI endoscopy. The cases also showed negative for parasite in stool examination. During the follow up period upper gastrointestinal endoscopy with biopsy from two to three areas, were performed which showed no eosinophilic infiltration.

Conclusion

Acute eosinophilic appendicitis is a rare event and less well understood entity and an early marker of acute appendicitis. Hence if patient undergoes laprotomy for various etiologies and if appendix found congested should be removed. Adopting such therapeutic modality can prevent future occurrence of acute appendicitis and hence need for appendectomy later in life. It is important to study AEA cases in detail for better understanding of disease pathogenesis and its significant role in patient management.

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