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RESEARCH ARTICLE

EOSINOPHILIC MYENTERIC GANGLIONITIS - A CAUSE OF MEGACOLON IN 3 DAY OLD GIRL: A RARE CASE REPORT

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Abstract

Eosinophilicmyentericganglionitis is an uncommon myenteric plexus inflammatory neuropathy with characteristic eosinophilic infiltration with or without hypoganglionosis. In our case, a full-term female newborn delivered via cesarean section had a delayed meconium transit, abdominal distension, and vomiting .Imaging findings showed the multiple dilated bowel loops. Microscopic analysis of the biopsy obtained from the rectosigmoid area shows presence of eosinophils in between the plexus fibres mainly around the ganglion cells.These features were consistent with the diagnosis of EosinophilicMyentericGanglionitis.

Introduction:-

Chronic intestinal pseudo-obstruction (CIPO) represents a rare and severe condition characterized by failure of the intestinal tract to propel its contents normally [1]. EMG is described under the broader classification of enteric ganglionitis. Myentericganglionitis is aptly named given the inflammatory involvement of Auerbachplexus[2].The condition presents with clinical features such as abdominal pain, vomiting, distended abdomen, constipation, and diarrhoea[1].The presentation of eosinophilicmyentericganglionitis (EMG) can be similar to that of Hirschsprung disease (HD)[3]. A high index of suspicion for EMG, albeit rare, should be entertained in cases of CIPO since these patients usually have normal colonoscopy and/or biopsies due to the involvement of the myenteric plexus and sparing of the submucosalplexus[4].

Case Report

A full term female baby delivered by lower segment cesarean section(LSCS) weighing 2.85 kg presented with adelayed meconium transit , abdominal distension , and vomiting. Mother was $G_2P_1L_1$ and delivered a female baby at 37 weeks. On physical examination,the baby had distension in the abdomen. On blood examination ,haemoglobin (10.8g/dl) ,total leucocyte count(2700 cells / cubic mm) and the platelet count was 15,000 cells/cubic mm. No eosinophilia was present in blood examination. RFT and LFT were within normal limits. On X-ray abdomen AP view, there was evidence of distended stomach and bowel loops. On USG abdomen, multiple dilated small bowel loops as well as large bowel loops were noted, the largest measuring 1.7 cm at jejunum and 2.2 cm at descending colon. On the basis of clinical and imaging findings, a diagnosis of Hirschsprung disease was made. The baby underwent surgery in which a segment of the dilated large bowel was resected and colostomy was done. Biopies from the rectosigmoid region and the colostomy site (transverse colon) were taken and sent for histopathological examination. On the gross examination, biopsy from the rectosigmoid region measured 0.4 x0.2 cm and biopsy from the colostomy site measured 0.5 x0.2 cm. On microscopic examination, biopsies from both the sites showed the presence of ganglion cells. There is a presence of eosinophils in between the plexus fibres, mainly

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around ganglion cells in the rectosigmoid biopsy. On the basis of histomorphological findings, the diagnosis of EosinophilicMyentericGanglionitis(EMG) was made.

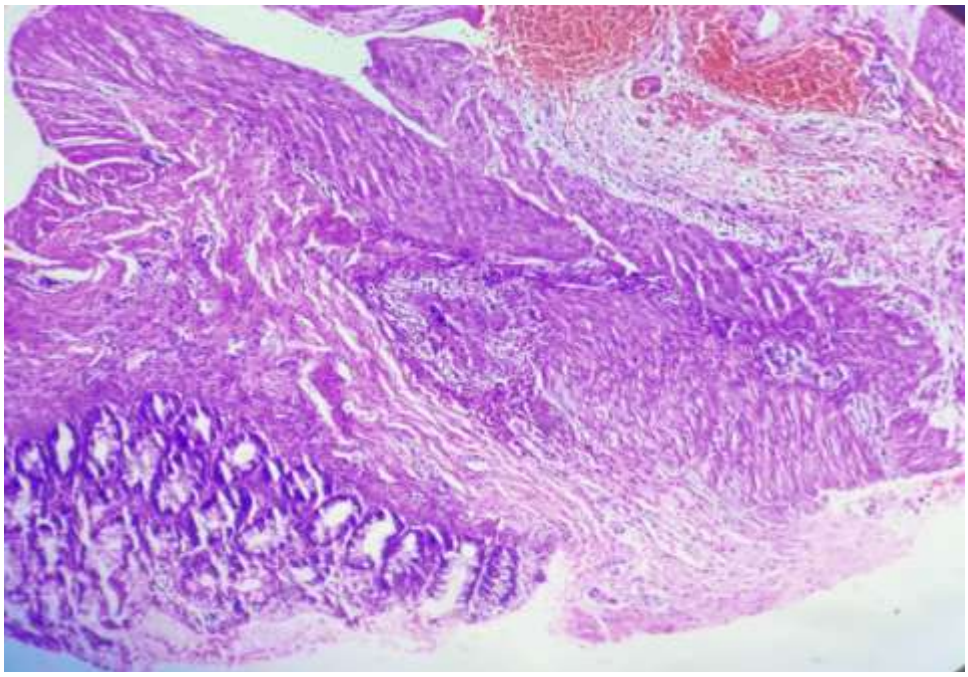

Figure 1:- Microscopy showing all the layers of colon (H & E stain, 100x).

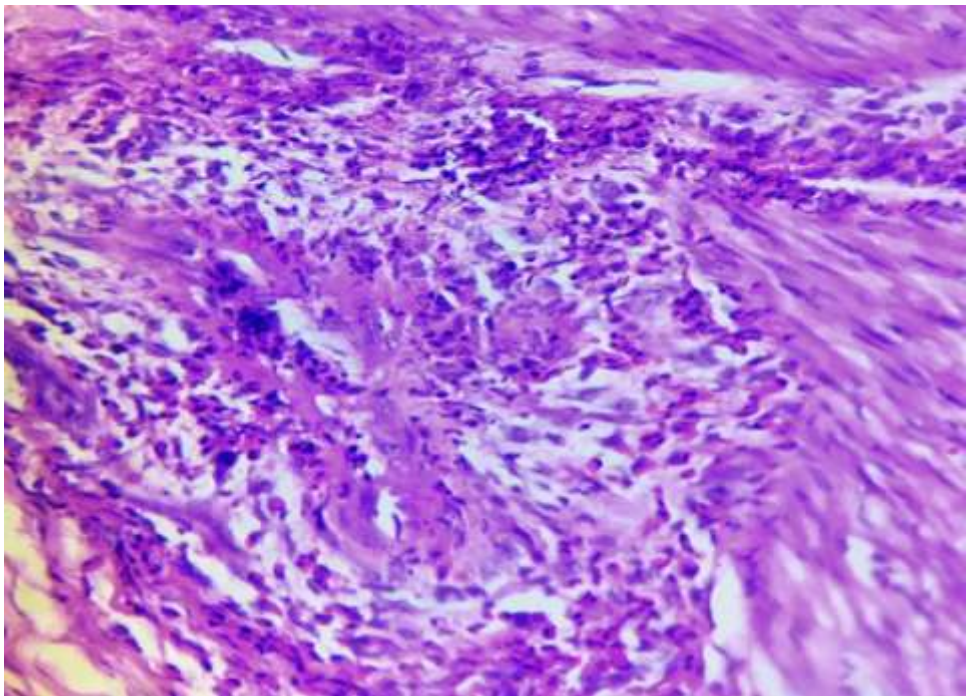

Figure2:- Microscopy showing myenteric plexus with eosinophilic infiltration between ganglion and glial cells. (H&E Stain, 400X).

Discussion:-

Eosinophilicmyentericganglionitis is characterized by eosinophilic infiltration of the Auerbach (myenteric) plexus, typically sparing the submucosa and mucosa, and is a rare cause of CIPO [4]. The disease is predominantly reported

in the paediatric population, with age of onset ranging from as early as infancy up to 15 years, our patient presented at birth[6] .The exact cause of the localized inflammation within the myenteric plexus in EMG remains unclear. Some authors suggest that the myenteric ganglia produce more IL-5, an eosinophil chemo- attractant, than surrounding regions. This localized inflammation may be due to an initial injury e.g., allergic, autoimmune, infection ,leading to Th2 cell infiltration of the region with subsequent increased IL-5 production and recruitment of eosinophils. Due to the over expression of IL-5, there is peripheral blood eosinophilia and compartmentalised distribution of eosinophils within the gastrointestinal tract but there is no peripheral eosinophilia in our patient. EMG diagnosis is made based on symptoms and histological examination results. Two-third of patients had symptoms, mainly diarrhoea and abdominal pain, poor feeding, vomiting and more rarely, as in this reported case, a pseudo-intestinal obstruction may occur[3]. Eosinophilicmyentericganglionitis is characterised by an absence of demonstrable lymphocytes within the inflamed enteric ganglia and is not associated with either morphological or functional evidence of enteric denervation, small intestinal phase III motor activity and reflex anorectal inhibitory responses are preserved in eosinophilicganglionitis[5]. The diagnosis requires histologic evaluation of the myenteric plexus, which cannot be done on routine mucosal biopsies due to their superficial nature. Thus, this diagnosis can only be made on resections(or rarely using laparoscopic full-thickness biopsies[2].

Conclusion:-

EMG is a rare condition presenting with intestinal pseudo-obstruction. Its clinical presentation resembles that of Hirschsprung disease. The final diagnosis requires full thickness biopsy from the colon.

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