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Case Report

Fatal Eosinophilic Gastroenterocolitis in a Two-Year-Old Child

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Summary. We report a case of eosinophilic gastroenterocolitis in a 2-year-old child with extensive fibrosis, atrophy of the muscularis propria and involvement of stomach, small bowel and colon. Following an attack of acute gastroenteritis at the age of 15 months the symptoms of ileus persisted. A biopsy of small bowel at the age of 18 months showed numerous eosinophilic granulocytes in the mucosa. At the age of 28 months the child died with paralytic ileus. This is the first case known to us of an eosinophilic gastroenteritis in early childhood with a fatal outcome.

Key words: Eosinophilic gastroenterocolitis – Case report – Occurrence in a 2-year-old child – Fatal outcome.

Introduction

Eosinophilic infiltration of the gastrointestinal tract (eosinophilic gastroenterocolitis) is an uncommon condition which is characterized by gastrointestinal symptoms, peripheral eosinophilia and a diffuse or circumscribed infiltration of the intestinal wall by mature eosinophils. When mucosal involvement predominates it results in protein-loss and malabsorption. Muscular involvement causes mainly obstructive symptoms, and predominantly subserosal changes lead to ascites with abundant eosinophils in the fluid (Klein et al., 1970).

The stomach is involved most frequently, twice as often as the duodenum, jejunum or ileum. The colon is rarely affected. Two cases of eosinophilic esophagitis have been reported (Landres et al., 1978; Dobbins et al., 1977). Among about 100 published cases with eosinophilic infiltration of the gastrointestinal tract there were 15 children, 5 cases showing the circumscribed variant (Samter et al., 1966; McGreevey et al., 1967; Toole et al., 1959; Welti et al., 1960) and 10 the diffuse variant (Klein et al., 1970; Jona et al., 1976; Katz et al., 1977).

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Here the case of a child is reported, who had persistent ileus symptomatic for 13 months following an attack of gastroenteritis.

Autopsy revealed scarring of the stomach and of both the small and large bowel. There was marked atrophy, especially of the outer muscle layer and little inflammatory activity was found at the time of autopsy.

Case Report

A 15 month old boy suffered from acute gastroenteritis with fever. After dietetic treatment in hospital and discharge when fever had disappeared, the diarrhoea became worse, and a laparatomy had to be performed one month later for intractable ileus. At laparatomy the small intestine, from the ligament of Treitz to the ileocoecal valve was markedly dilated without a recognizable cause for mechanical ileus. Roentgenologically several fluid levels were seen in the small bowel, and two weeks later a second laparatomy was necessary; however, no morphological cause for the ileus symptomatic was found. Blood examination revealed a leucocytosis of 20,000 cells and a leftshift, with an eosinophilia of about 16%. A Coli-, Klebsiella and Candida sepsis occured. Parts of the small bowel were gas filled and showed changes resembling megacolon after electric stimulation and mechanical stress. At a third laparotomy two weeks later an ileostomy in the distal ileum was carried out. Intraoperatively a biopsy of the small bowel, the ileostomy and the liver was performed.

The small intestine showed a highly cellular granulation tissue with numerous eosinophils, mainly within the mucosa and submucosa (Fig. 1a and b). The liver biopsy revealed groups of eosinophils within the sinusoids (Fig. 1c) and the diagnosis of an eosinophilic enteritis was made. In the biopsy of the ileostomy, part of the normal myenteric plexus could be seen. Despite many therapeutic efforts, no peristaltic waves were observed. The child died at the 20. 10. 1978 at the age of 28 months, after thrombosis of the vena cava.

Autopsy (SN 696/78) revealed partially dilated small bowel and colon with thinned-out intestinal wall. Histologically there was extensive fibrosis of the submucosa and serosa of the stomach, and of the small bowel and colon, with a partially interrupted muscularis mucosae and severe atrophy of the outer layer of the muscularis propria (Fig. 2a and b). Low grade chronic inflammation of the mucosae of the entire intestinal tract was seen. The submucous and myenteric plexuses appeared to be normal. Eosinophilic infiltration was not present in tissues examined post-mortem. There was a low grade, older, fibrinous peritonitis in the epigastrium. The pancreas was fibrotic and fibrosis involved the urinary bladder and the ureters with low grade hydroureter and hydronephrosis. Old thrombosis of the superior vena cava extended from the right cardiac atrium to both jugular veins. The inferior vena cava also contained an old thrombus extending from the right renal vein to the hepatic vein. Fatty liver, ascending pyelonephritis and chronic bronchitis were also found.

Discussion

Eosinophilic leucocytes are normal components of gastrointestinal tissue. An increase in number is found in such diverse pathologic states as Hodgkin's

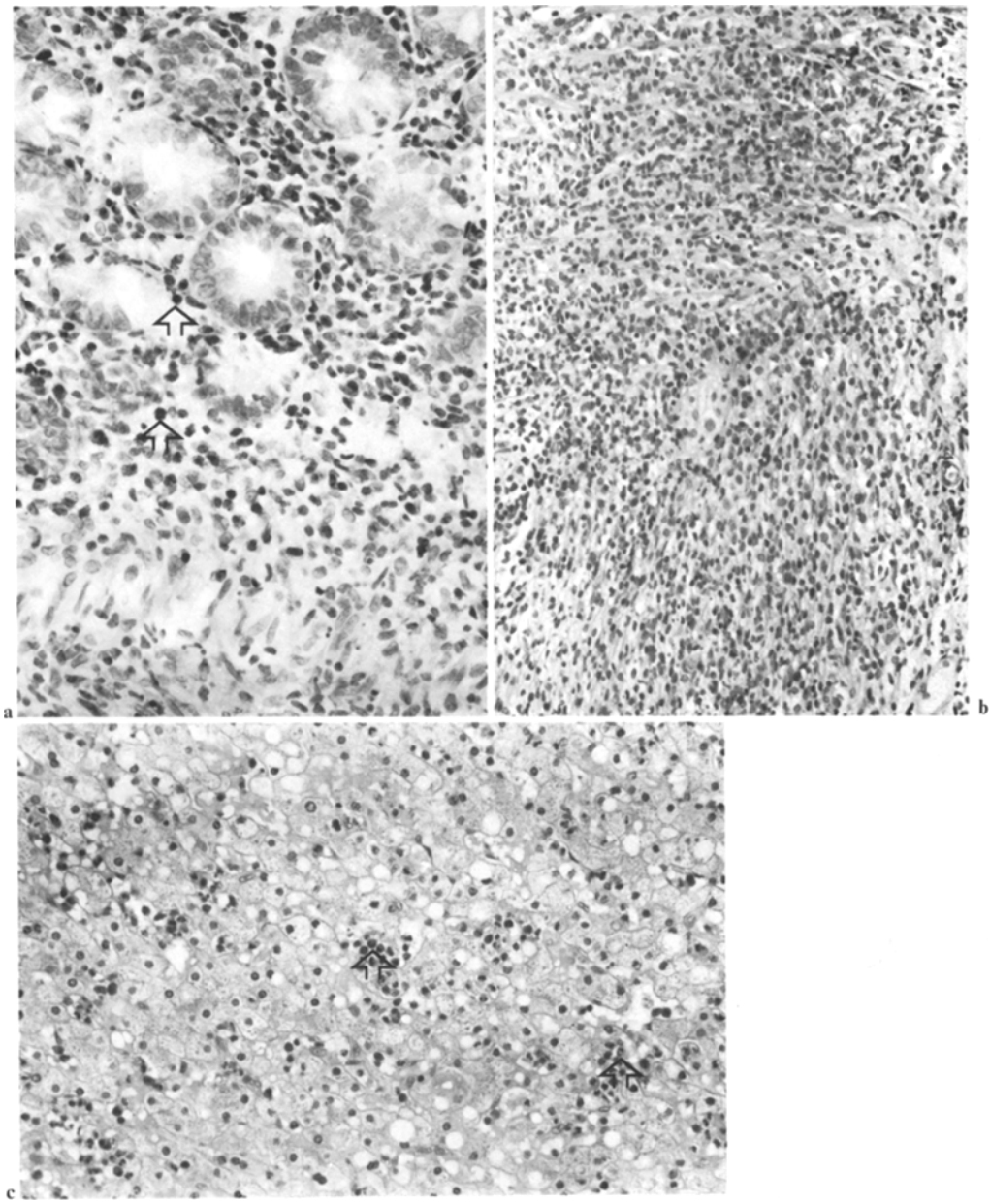
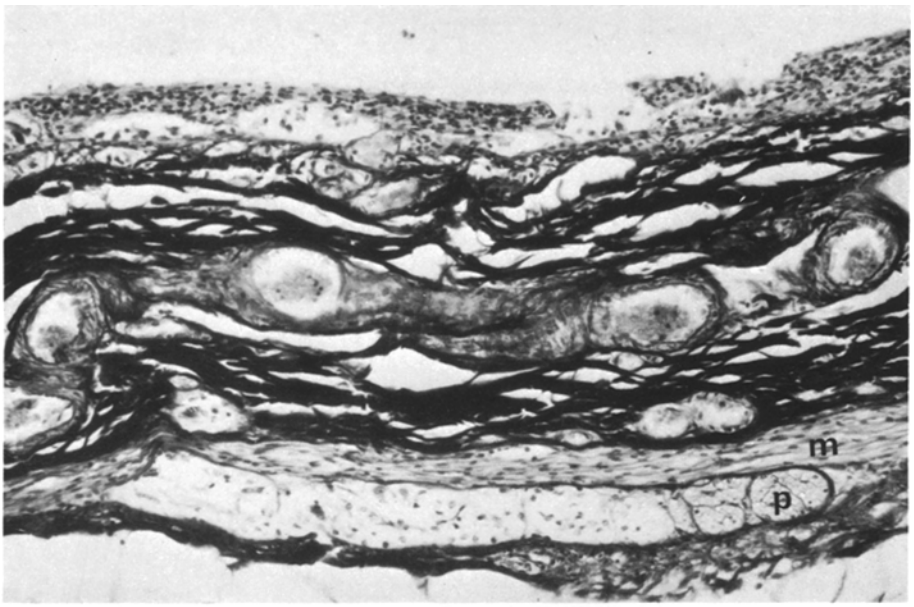
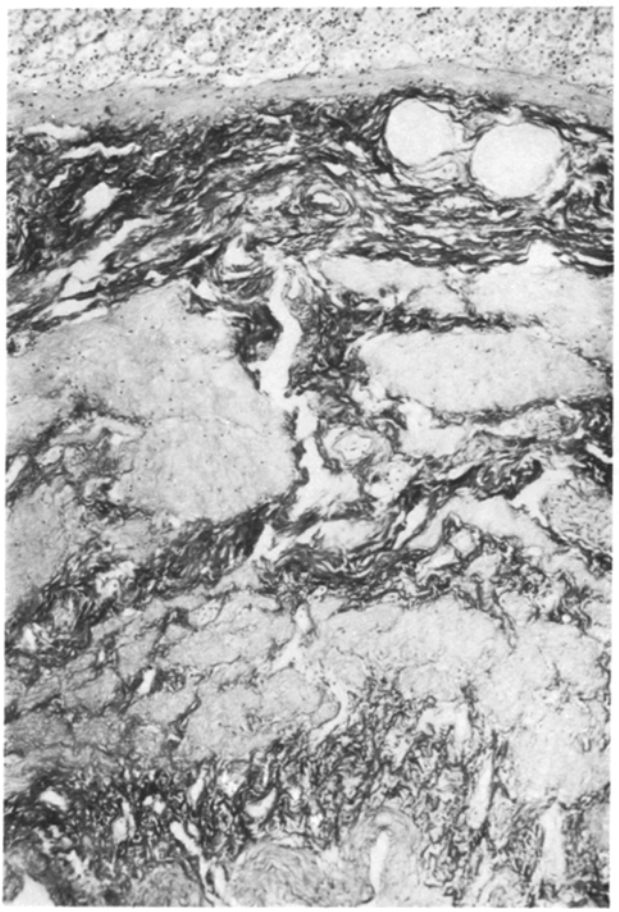


Fig. 1. a Eosinophilic enteritis: biopsy of small bowel mucosa shows cellular infiltration with predominantly eosinophils (dark nuclei; ⇒). Biebrich scarlet red, × 250. **b** Eosinophilic enteritis: granulation tissue showing diffuse inflammatory infiltration of the submucosa of small bowel. Numerous eosinophils with dark nuclei. Biebrich scarlet red, × 160. **c** Liverbiopsy with numerous eosinophils (⇒), predominantly in the liver sinus. Biebrich scarlet red, × 160



a



b

Fig. 2. **a** Colon: excessive fibrosis of submucosa and atrophy of the inner (*m*) and outer muscle layer with normal plexus myentericus (*p*) between the two muscle layers. elvG, ×100. **b** Extensiv fibrosis of submucosa and subserosa of the stomach. elvG, ×63

disease, gastric ulcer, gastrointestinal carcinoma, the hypereosinophilic syndrome and disseminated eosinophilic collagen diseases. Various terms have been used for the disease entity known as eosinophilic gastroenteritis: pyloric hypertrophy with eosinophilic infiltration, the gastric lesion of Loeffler's syndrome, gastric granuloma with eosinophilic infiltration, eosinophilic granuloma or infiltrative eosinophilic gastritis.

Morson and Dawson (1972) distinguished three groups of eosinophilic infiltrative or granulomatous lesions of the gastrointestinal tract. In eosinophilic gastroenteritis there are single or multiple foci with thickening of the pylorus or variably long segments of intestine, which can cause obstructive symptoms. There is usually a blood eosinophilia, occasionally a malabsorption or protein-loosing enteropathy. In about half of the cases there is an allergic diathesis.

Macroscopically the affected segment is oedematous and thickened with narrowing of the lumen. Histologically abundant eosinophils are found, especially in the lamina submucosa and sometimes also in the muscularis propria or serosa. Fibrosis of the submucosa in this group is not marked. The second group, the eosinophilic granulomatous polyp or inflammatory fibroid polyp, usually presents as a localized lesion without eosinophilia in the blood. The polyps are usually single measuring up to 4 cm in diameter, arising in the submucosa. The most common symptom is acute intestinal obstruction with intussusception. The third group, allergic gastroenteropathy, is found in children and is characterized by anemia, oedema, growth retardation, hypo-albuminaemia, hypogammaglobulinaemia, blood eosinophilia and excessive gastrointestinal protein-loss (Waldmann et al., 1967). Obstruction and macroscopically identificable local lesions are uncommon; histologically little eosinophilic infiltration in the lamina propria is found.

The case presented resembles the third group of "eosinophilic gastroenteritis". It shows an extraordinarily diffuse infiltration of stomach, small bowel and colon and severe fibrosis of the submucosa and muscularis propria, marked atrophy of the outer muscle layer and a partially interrupted muscularis mucosae. This is in contrast to other published cases, which show muscular hypertrophy with involvement of the muscular layer. Necrosis of muscle fibres is mentioned occasionally (Johnstone and Morson, 1978). At autopsy scarring of the intestinal wall was found, with little residual inflammatory activity and few eosinophils to be seen. In our case, operative liver biopsy had shown eosinophils within sinusoid.

An eosinophilic cystitis has been reported in association with eosinophilic gastroenteritis in three patients (Palubinskas, 1960; Gregg and Litz, 1974). It is possible that in our case the fibrosis of the urinary bladder and ureters may be sequelae of an eosinophilic cystitis and uretitis.

Reviewing the clinical data of eosinophilic gastroenteritis in childhood the duration of symptoms is seen to vary from several hours to 7 years (Table 1). This is the first case of an eosinophilic gastroenterocolitis with fatal outcome in this age group. Previously there has been only one report of death in an adult, a 66-years-old man with eosinophilic enteritis of the jejunum (Tytgat et al., 1976).

Table 1. Summary of pediatric patients with diffuse eosinophilic infiltration of the gastrointestinal tract

Author	Age, sex	Symptoms	Duration	Clinical presentation	Eosinophilia	Pathologic-anatomic location	Operation	Present state
Klein et al.	16, ♂	fatigue, bile regurgitation	?	antral disease	10%	antrum duodenum jejunum	partial gastrectomy	well
Jona et al.	11, ♂	weight loss fatigue anemia	3 months	antral disease	4%	antrum appendix	antral biopsy appendectomy	well
Jona et al.	12, ♀	mild abdominal pain	3 days	acute appendicitis	11%	appendix	appendectomy	?
Jona et al.	6, ♀	severe abdominal pain	several hours	acute appendicitis	2%	appendix	appendectomy	well
Katz et al.	10, ♂	growth retardation abdominal pain vomiting	2 years	small intestine	0%	stomach small intestine	biopsy of stomach and small intest.	well
Katz et al.	8, ♀	adominal pain asthma	7 years	small intestine	2%	stomach small intestine	small intest.	well
Katz et al.	$3^1/_2$, ♂	growth retardation chronic diarrhoea	$8^1/_2$ years	small intestine	0–2%	stomach small intestine	small intest.	well
Katz et al.	2, ♂	growth retardation asthma	$4^2/_3$ years	small intestine	3%	stomach small intestine	small intest.	well
Katz et al.	$^1/_2$, ♂	growth retardation chronic diarrhoea	$^1/_2$ year	small intestine	0–3%	stomach small intestine	small intest.	well
Katz et al.	$^1/_2$, ♂	anemia cardiac failure	$^1/_2$ year	small intestine	2%	stomach small intestine	small intest.	well
Present report	$1^1/_4$, ♂	diarrhoea ileus	13 months	small intestine colon	16%	colon small intestine colon	biopsy of small bowel	letal

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