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Case Report

Dysphagia as Presenting Symptom of Crohn's Disease in a 13-Year Old Girl

Berbel Kooger[1]*, Prapto Sastrowijoto[2], Anita Van den Neucker[3], Eric Keulen[4], and Philippe Rosias[1]

[1]*Department of Pediatrics, Zuyderland Medical Center, Sittard-Geleen and Heerlen, Netherlands*

[2]*Department of Pathology, Zuyderland Medical Center, Sittard-Geleen and Heerlen, Netherlands*

[3]*Department of Pediatric Gastroenterology, Maastricht University Medical Center, Maastricht, The Netherlands*

[4]*Department of Gastroenterology, Zuyderland Medical Centre, Sittard-Geleen and Heerlen, Netherlands*

***Corresponding author**

Berbel Kooger, Department of Pediatrics, Zuyderland Medical Center, Sittard-Geleen and Heerlen, Dr. H. van der Hoffplein 1, 6162 BG, Geleen, Netherlands, Tel: 3188-459-7777; Email: berbelkooger@gmail.com

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Abstract

Esophageal Crohn's disease (CD) is rare. We present a young girl with dysphagia, weight loss and retrosternal pain due to esophageal CD. Diagnosis was only confirmed after repeated esophageal biopsies and an additional ileocolonoscopy. Abdominal ultrasound and fecal calprotectin did support the diagnosis of CD. Treatment by exclusive enteral feeding, temporary oral viscous budesonide, esomeprazole, and azathioprin resulted in gradual remission of ulcerations. Due to loss of response after six months, maintenance therapy was switched into methatrexat and subsequently into infliximab. In literature, only a few cases have been reported. Both esophagogastro- and ileocolonoscopy should be considered in case of aspecific upper gastrointestinal symptoms and despite absent lower gastrointestinal symptoms.

ABBREVIATIONS

CD: Crohn's disease; GI: gastrointestinal

INTRODUCTION

Crohn's disease (CD) is an idiopathic chronic inflammatory disorder of the gastrointestinal tract. It is characterized by chronic ulcerative transmural inflammation, and may be localized throughout the digestive tract. Most frequently, lesions are localized in the terminal ileum and colon. Furthermore, fistula, stenosis, and strictures are common. Prevalence of pediatric CD in the United States is 43 per 100.000 children, whereas exact prevalence in Europe is unknown. Incidence of CD in children under 18 years in the Netherlands was from 1999 until 2001 5.2 per 10^5 per year [7]. Children often suffer from abdominal pain, bloody diarrhea and weight loss. Upper gastrointestinal (GI) involvement is less common (30% of children). Esophageal CD is rare (1-10%, [6]). We report a case of a young teenager with esophageal involvement, which initially presented with dysphagia, weight loss and retrosternal pain.

CASE PRESENTATION

A previously healthy 13-year old girl presented with complaints of progressive dysphagia and extreme retrosternal pain since 3 weeks. Chewing was normal. She experienced discomfort with swallowing of both liquid and solid food. She also experienced pyrosis and a globus sensation. Her appetite was decreased and she lost 3 kilogram. Analgetics had no effect. There was no history of abdominal complaints, fever, bloody stools, vomiting, hematochezia, melena, diarrhea, drug abuse, medication use or caustic ingestion. Family history was negative for inflammatory bowel disease (IBD) and autoimmune disease.

Physical examination was normal. No oral aphthous lesions and no erythema nodosum were seen on inspection. Weight and height were respectively +0.1SD and -0.1SD at 10 years age. Biometry and relevant laboratory results are shown in Table (1). An X-ray to examine passage of esophagus-stomach-duodenum showed normal esophageal motility and no evidence for obstruction. Stool cultures were negative for viruses, bacteria, and parasites.

An esophagogastroduodenoscopy showed macroscopic punched out ulcers of 3-5 mm depth, linearly ranked, continuously beyond Z-line, without stenosis or fistula (Figure 1,2). Histology demonstrated chronic active ulcerative, non-granulomatous inflammation. Furthermore, biopsies did not reveal evidence

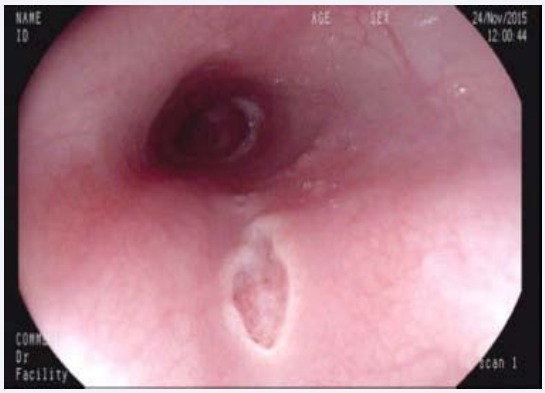

Figure 1 Deep esophageal ulcerations (3-5 mm).

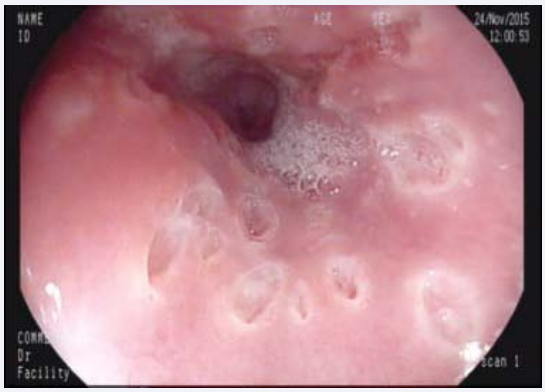

Figure 2 Deep esophageal ulcerations (3-5 mm).

for viral infection, fungal disease, caustic injury, collagenic or eosinophilic esophagitis.

Exclusive enteral tube feeding for sixteen days, and antacid and morphine did not result in improvement. To extend the diagnosis combined esophagogastroduodenoscopy and ileocolonoscopy was performed. Macroscopic esophageal findings did not differ. Microscopy demonstrated chronic active ulcerative granulomatous transmural extended inflammation (Figure 3).

Furthermore, terminal ileitis and right hemicolitis were found by ileocolonoscopy. Colonic biopsies showed chronic active ulcerative granulomatous inflammation matching CD. Fecal calprotectin was elevated (1800 µg/g). Abdominal ultrasound showed bowel wall thickness and stiffness and perienteric fluid. Patient was treated with exclusive enteral feeding, temporary viscous oral budesonide, esomeprazole, and azathioprin was started as maintenance therapy. In consultation with parents and the pediatric gastroenterologist, we initially followed this therapeutic approach. Six weeks after diagnosis of CD, patient experienced bloody stools. Ulcerative esophageal lesions recovered after approximately three months, proven by esophagogastroduodenoscopy. Six months after diagnosis, loss of response occurred under azathioprin, and maintenance therapy was switched into methotrexate. Unfortunately, the patient had loss of control with nausea and abdominal pain. Therefore, maintenance therapy was switched into infliximab.

DISCUSSION

CD is characterized by chronic, granulomatous, transmural inflammation of the complete gastrointestinal tract. Typically, esophageal CD presents with dysphagia, odynophagia, weight loss, pyrosis, retrosternal pain, nausea, and vomiting [2,3,8], although esophageal lesions may also be asymptomatic [4]. Specific endoscopic findings for esophageal CD comprise of aphthous ulcers, deep ulcerations, cobblestone appearance, erythema, pseudopolyps, stricture and fistula [2,3,5]. Histological findings include acute or chronic inflammation, ulceration and granulomas [3]. Diagnosis of esophageal CD is difficult, in particular in the absence of typical intestinal symptoms [4]. In general, other causes such as reflux esophagitis, viral/eosinophilic esophagitis, carcinoma, drug-induced and corrosive ulceration must be excluded. The presence of endoscopic esophageal ulcers is suggestive for esophageal CD [5]. Only a few case reports of isolated esophageal involvement are reported in literature [2]. In a large cohort (N=210) of pediatric CD patients all children with esophageal CD also had intestinal CD, whereas no children with isolated esophageal CD were identified [4]. In the presented case, the patient developed lower GI tract symptoms only six weeks after diagnosis. However, ileocaecal lesion could have already been present without clinical symptoms.

First esophageal biopsies only showed aspecific chronic ulcerative non-granulomatous inflammation. However, two weeks later, both esophageal and ileocolonic biopsies showed chronic ulcerative transmural inflammation with presence of granulomas. Granulomas are not always obvious [1] and biopsy specimens are often non-specific [6]. In a retrospective observational study (N=24) with patients with esophageal CD, granulomas were rarely seen [8]. Typical noncaseating granulomas occur in only less than 25% of reported cases [1]. Granulomas in CD are most likely located in the deep submucosa and lamina muscularis [1]. Microscopic chronic inflammation seen in esophageal biopsies may be suggestive for CD [5]. Histology is mostly supportive but not specific for CD [8].

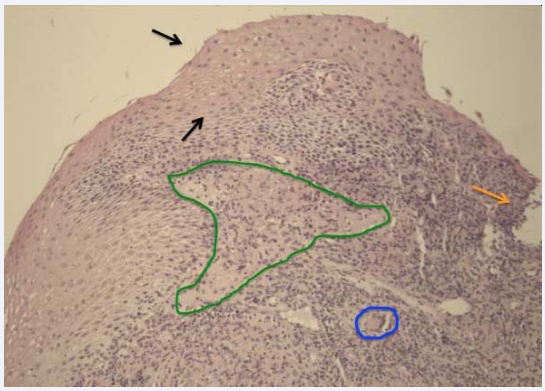

Figure 3 Histology of the esophagus with a giant cell (blue circle) and epitheloid and histiocytes (green area). Black arrow: normal mucosa. Orange arrow: ulcerative mucosa.

Table 1: Laboratory tests.

Variable	Value	Normal range	Variable	Value	Normal range
Weight	45.5 kg (-0.5 SD)	-	Folic acid	16.7 nmol/L	2.7-16.3 nmol/L
Height	158.3 cm (-0.7SD)	-	ALAT	20 U/L	< 44 U/L
Body mass index (BMI)	18.2 kg/m^2 (-0.1SD)	-	ASAT	17 U/L	< 48 U/L
Hemoglobin[a]	8.5 mmol/L	7.1-9.04 mmol/L	Bilirubin total	4.0 µmol/L	3-20 µmol/L
White blood cell count	7.8 x 10^9/L	4.0-14.0 x 10^9/L	Creatinin	59 µmol/L	50-71 µmol/L
Neutrophils	65%		Urea	4.6 mmol/L	2.6-6.8 mmol/L
Thrombocytes	345 x 10^9/L	150-350 x 10^9/L	25(OH)D$_3$	57 nmol/L	25-109.8 nmol/L
Ferritin	45 µg/L	13.7-78.8 µg/L	IgE serum	64.8 kU/L	9-407 kU/L
C-reactive proteine	22 mg/L	< 1 mg/L	tTG IgA	<1 kU/L	<1 kU/L
Erythrocyte sedimentation rate	38 mm/hour	< 20 mm/hour	IgA	1.9 g/L	0.47-2.2 g/L
Albumin	36 g/L	36.6-47.6 g/L	ANA & ANCA	Negative	Negative

a) With normal red blood cell indices

Pediatric CD is treated by exclusive enteral feeding. Subsequently, azathioprin may be started as maintenance therapy. In case of no remission, azathioprin should be switched to methotrexate or infliximab. The presence and extensiveness of upper gastrointestinal CD is important in the intestinal CD disease progression and recurrence. Probably, early therapy with biological could lead to less complication (stricture, fistula, and abscess) and prevent disease progression [8]. In case of strictures or stenoses, surgery may be needed.

CONCLUSION

In conclusion, esophageal CD is rare. Diagnosis of esophageal CD is difficult in absence of typical intestinal symptoms. Misleading by initial biopsies is demonstrated in this case, and could lead to under diagnosis, due to the lack of specific histological findings in the mucosa. In case of isolated aspecific upper gastrointestinal symptoms, both gastro and ileocolonoscopy should be considered.

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