Bleeding from duodenal lymphangiectasia

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Abstract

An 8 year old girl with recurrent upper gastrointestinal bleeding was found to have localised duodenal lymphangiectasia by fibre-optic endoscopy. She did not show physical signs or laboratory evidence of significant enteric protein loss. A low fat diet seemed to prevent further bleeding. Duodenal lymphangiectasia may be associated with gastrointestinal bleeding in children.

Intestinal lymphangiectasia is a disease characterised by dilated lymphatic channels in the mucosa, submucosa, and serosa, as well as mesentery of the intestine.¹ This generalised disorder of lymphatics usually occurs in the small intestine and causes excessive intestinal protein loss.¹ The variant of intestinal lymphangiectasia in which blood, as well as protein, was lost in the bowel was described in two adult patients.^{2 3}

We report a case of a girl with repeated upper gastrointestinal bleeding associated with duodenal lymphangiectasia without enteric protein loss.

Case report

An 8 year old girl was referred for further investigation of her upper gastrointestinal bleeding. Two days before admission she experienced painless haematemesis and melaena. Her parents denied any kind of previous drug ingestion, nose bleeding, diarrhoea, vomiting, or retching.

On admission the clinical findings were normal. Her body weight was 27 kg and height 126 cm; these were both on the 50th centile.

Laboratory investigations showed a haemoglobin concentration of 90 g/l, and a leucocyte count of $9.4 \times 10^9/l$ with lymphocytes at $4.5 \times 10^9/l$. Her erythrocyte sedimentation rate was 15 mm in the first hour. Urinalysis was normal. Total serum protein concentration was 58 g/l and albumin 36 g/l. Repeated stool examination for ova and protozoa was negative. Contrast radiography of the upper intestinal tract was normal. Abdominal technetium scanning was negative. Ear, nose, and throat examination and laryngotracheoscopy were negative.

After two days of fasting upper gastrointestinal endoscopy showed a normal oesophagus, stomach, and duodenal bulb. Descending duodenal mucosal folds were oedematous with

(A) Duodenal biopsy: villi are almost completely occupied by a considerably dilated lymph vessel (*). In the lymphangiectasia proteinaceous material is visible. (B) and (C) Duodenal endoscopic pictures: ridge like mucosal projections covered by milky material (chyle) (▲) and numerous white elevations of lymphangiectasia (◊) are prominent.

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numerous whitish elevations, each the size of a millet seed (figure B and C). Multiple mucosa biopsy specimens demonstrated intestinal lymphangiectasia. Duodenal villi were almost completely occupied by considerably dilated lymph vessels (figure A). In the lymphangiectasia a chyle like substance was visible.

After this finding additional examinations were made: faecal fat and serum immunoglobulin concentrations, α_1 antitrypsin activity, and the CD4:CD8 T cell ratio were all within normal limits. Faecal α_1 antitrypsin clearance was not examined.

The girl was discharged with recommendations of dietary fat reduction and iron supplementation. She did not attend outpatient follow up.

Ten months later she experienced another similar attack of upper gastrointestinal bleeding. She had followed the dietary recommendations for only two months. At admission her haemoglobin concentration was 85 g/l, leucocyte count $10.5 \times 10^9/l$ with lymphocytes at $3.8 \times 10^9/l$. Total serum protein and albumin and serum immunoglobulin concentrations, prothrombin time, coagulation factors, serum glutamic oxaloacetic transaminase and glutamic pyruvic transaminase activities, faecal fat excretion, blood glucose, and blood urea nitrogen were within normal limits.

Repeated upper gastrointestinal contrast radiography and abdominal technetium scanning were negative. Ear, nose, and throat examination and laryngotracheoscopy were normal.

Fibreoptic examination of the oesophagus, stomach, and duodenal bulb did not show any bleeding lesion. In the descending duodenum a few streaks of fresh blood were visible. On histological examination mucosal changes of lymphangiectasia were prominent.

She was discharged with identical dietary recommendations and medical treatment. During four years' follow up she has not experienced haematemesis and/or melaena and her blood count remains normal. Further fibreoptic duodenoscopy demonstrated less pronounced mucosal fold thickening and fewer whitish spotty mucosal elevations.

Discussion

Intestinal lymphangiectasia in the paediatric age group may vary widely in its manifestation and severity.¹ It usually presents with chronic diarrhoea with protein loss and growth retardation.¹ Treatment of intestinal lymphangiectasia with a high protein, fat free diet with added medium chain triglyceride is usually effective in preventing or alleviating the

diarrhoea and hypoproteinaemia.¹ The absence of fat in the diet prevents engorgement of the intestinal lymphatics with chyle, thus preventing their rupture with its concomitant protein loss.¹ The intestinal lesion may be patchy in distribution and more or less localised.^{1 4 5} Thus clinical presentation and symptom severity may depend on the anatomical location and extent of this anomaly.^{1 5} Intestinal lymphangiectasia may be asymptomatic or symptoms may be provoked by an exogenous trigger—for example, fatty meals.⁶

The present case seems to represent a particular variant of intestinal lymphangiectasia in which gastrointestinal blood losses occurred in the absence of gross protein and fat loss due to short segmental involvement. The striking association between apparent gastrointestinal bleeding and duodenal lymphangiectasia contrast with the opinion that the only connection between the lymphatic and systemic circulation are where the large thoracic and right lymphatic ducts join the great veins. The existence of latent lymphatic-venous anastomosis and lymphatic-arterial communications that can be opened under certain conditions has been already demonstrated.^{2 3} An obstruction of the normal efferent flow of the chyle from the intestine may in turn raise the lymphatic pressure to a level sufficiently high to open latent lymphatic-venous or abnormal lymphatic-arterial connections.^{2 3} This allows retrograde flow of blood into the lymphatics from the system of a higher pressure.^{2 3} Thus the rupture of thin walled and blood filled dilated mucosal lymphatics into the duodenum as a result of the increased pressure within them may give rise to symptoms of intestinal bleeding.

In conclusion, in some children with proximal gastrointestinal haemorrhage duodenoscopy and mucosal biopsies may show a particular association between intestinal bleeding and lymphangiectasia. In our case it seems that dietary manipulation may have had an influence on the course of disease.

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- 1 Vardy PA, Lebenthal E, Shwachman H. Intestinal lymphangiectasia: a reappraisal. *Pediatrics* 1975;55:842–51.
- 2 Davidson JD, Flynn EP, Kirkpatrick JB. Protein-losing enteropathy and intestinal bleeding. The role of lymphatic-venous connections. *Ann Intern Med* 1966;64:628–35.
- 3 Poirier VC, Alfidi RJ. Intestinal lymphangiectasia associated with fatal gastrointestinal bleeding. *Dig Dis Sci* 1973;18:54–8.
- 4 Abramowsky C, Hupertz V, Kilbridge P, Czinn S. Intestinal lymphangiectasia in children: A study of upper gastrointestinal endoscopic biopsies. *Pediatr Pathol* 1989;9:289–97.
- 5 Van der Meer SB, Forget PP, Willebrand D. Intestinal lymphangiectasia without protein loss in a child with abdominal pain. *J Pediatr Gastroenterol Nutr* 1990;10:246–8.
- 6 Veldhuyzen van Zanten SJO, Bertlsman GNJ, Tygat GNJ. Endoscopic diagnosis of primary intestinal lymphangiectasia using a high-fat meal. *Endoscopy* 1986;18:108–10.