Angiodysplasia and ileal carcinoid

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Abstract

A case of angiodysplasia, symptomatic for 18 years, is presented. This case highlights the difficulty of establishing a diagnosis. Associated findings included aortic stenosis and a carcinoid tumour of the ileum. Despite transfusion of a total of 1200 units of blood the liver was normal at necropsy.

Case report

A 60 year old man presented in 1971 with iron deficiency anaemia. Past history included a sigmoid colectomy for adenocarcinoma. Significant clinical findings included angiomas on the undersurface of the tongue, the murmur of aortic stenosis (confirmed by cardiac catheterisation), and episodic melaena. Gastrointestinal tract bleeding continued for a further 18 years until his death, by which time he had required a cumulative transfusion of approximately 1200 units of blood.

Angiodysplasia had been suspected clinically from the outset. Many investigations had been performed to locate the site of bleeding in the gastrointestinal tract. These included multiple endoscopies of the upper and lower gastrointestinal tract, barium studies, abdominal ultrasonography, radiolabelled isotope scans, and selective arteriographic studies during bleeding episodes, and two laparotomies. Angiodysplasia could not be confirmed during life.

In 1980 during one episode of active bleeding hypotonic duodenography was performed. An area of angiodysplasia was suspected in the distal duodenum/proximal jejunum. Jejunal resection (90 cm) was performed to include this area. The specimen was grossly normal and routine histological examination was normal. Blood loss stopped for three months.

Thereafter bleeding recurred and monthly transfusions were required. Hospital admissions became more frequent and prolonged. In 1988 telangiectasia was noted in the distal duodenum at endoscopy. The patient declined further investigations or surgery and died one year later.

At necropsy the stomach and bowel were removed intact from the lower oesophagus to anus. The major vessels, the coeliac axis and the superior and inferior mesenteric arteries were injected with saline and then with contrast agent. An arteriogram was performed which showed no appreciable abnormality. The lumen of the gastrointestinal tract was flushed through with saline and then filled with formalin for three days to obtain maximum fixation.

Three days later the bowel was opened, pinned out, and allowed to fix further in formalin. It was subsequently examined macroscopically and with a dissecting microscope. No abnormality was detected. Sections from the entire stomach and intestine were 'swiss rolled' (approximately

100 blocks). All sections of the entire stomach and small and large bowel were examined histologically.

Two discrete areas of vascular dilatation, consistent with a diagnosis of angiodysplasia, were found microscopically, one in the stomach at the oesophagogastric junction and the other in the small bowel approximately 50 cm from the gastroduodenal junction. The vascular lesions were characterised by the presence of dilated vascular channels in the submucosa extending into the mucosa. The maximum size was 2 mm. Examination of the anastomotic site of the previous resection showed no evidence of carcinoma.

Further findings at necropsy included an 0.8 cm carcinoid tumour of the terminal ileum (Figure). The regional lymph nodes were not involved. Aortic stenosis was confirmed. In addition, vegetations composed of fibrin and Gram positive cocci were found on the aortic and tricuspid valves; a diagnosis of infective endocarditis secondary to a longterm central line inserted for venous access was made. A splenic infarct was seen, presumably related to embolisation from the infective endocarditis. There was no evidence of chronic hepatitis or of increased iron deposition in the liver or in the spleen. There was no evidence of haemosiderosis,

The carcinoid tumour is composed of nests and tubules lined by small cells with hyperchromatic nuclei and abundant eosinophilic cytoplasm.

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which is interesting in view of the massive cumulative transfusion.

Discussion

Angiodysplasia, first described by Galdabine in 1974, is characterised by microvascular abnormalities of the mucosa and submucosa of the gastrointestinal tract. They are frequently multiple and are most commonly found in the caecum and ascending colon. The arterioles typically show a normal appearance,¹ and are often limited to small areas of 5 to 10 mm. They may be found in the presence or absence of other disease of the gastrointestinal tract such as ulcerative colitis and neoplasia. While the appearances of the vascular channels are not specific, in view of their site and multiplicity the diagnosis of gastrointestinal vascular malformations/angiodysplasia may be made.

Angiodysplasia, which has been divided into three types on the basis of clinical features by Moore *et al.*,² may represent the most common cause of obscure gastrointestinal bleeding in the elderly population.³ While the caecum is said to be the predominant site it is being increasingly recognised in the stomach^{4,5} and small bowel.^{6,7} Fibreoptic endoscopy remains the most useful method of diagnosis of mucosal lesions in the stomach and large bowel. For mucosal and submucosal lesions in the small bowel, mesenteric angiography is the most useful diagnostic technique. The latter, however, is only useful if the lesions are actively bleeding at the time of the study. Vascular tufts seen in the arterial phase and early filling of the draining vein and slow emptying of dilated intramural veins are the characteristic features of angiodysplasia. Some authorities have found improved localisation of the lesions if the colon is distended with air at the time of the study. Direct serial magnification radiography may be used at the same time.⁸

Even in the resected specimen angiodysplasia may be missed by naked eye inspection and routine histological analysis. A detailed microscopic analysis of the entire gastrointestinal tract of this patient showed only two small lesions at necropsy.

The association of vascular malformations with other diseases has been reported, including aortic stenosis, chronic renal failure,⁹ liver cirrhosis,¹⁰ and many other systemic illnesses. The association in this case with a primary carcinoid tumour of the small bowel is previously unreported. Carcinoid tumours account for less than 1% of all tumours of the gastrointestinal

tract and of these, approximately 15% occur in the ileum. They arise from neuroendocrine cells and frequently secrete certain amines and peptides. Bleeding may occur as a result of peptic ulceration particularly due to gastric and duodenal carcinoids, though this was not the case in our patient.

Angiodysplasia is difficult to treat. Several options are available including simple supportive treatment for anaemia, surgical resection, endoscopic electrocautery,¹¹ selective embolisation of the gastroduodenal artery,¹² sclerotherapy, and laser¹³ and hormonal therapy.¹⁴ Many of these are of limited value because of the multifocal and varied position of the lesions. In many instances repeated transfusion is required, as in this case.

We have reported the case of a patient with an 18 year history of gastrointestinal tract bleeding due to angiodysplasia, requiring 1200 units of blood. Despite the use of sophisticated radiology, the diagnosis could not be confirmed during life. In addition, the patient also had had an asymptomatic carcinoid tumour of the small bowel.

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