A more lymph invasive tumour compared with those with HALN negative liver metastases.

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An unusual case of colonic angiodysplasia

N J Trendell-Smith, B F Warren, E A Sheffield, P Durdey

Abstract

An unusual case of a colonic vascular anomaly resembling angiodysplasia associated with right sided diverticular disease is presented. The patient, a 74 year old man, presented with a four day history of rectal bleeding and subsequently underwent hemicolectomy. The resected specimen was flushed out with heparin–saline solution and injected with a barium–gelatine mixture. Preoperative barium enema revealed right sided diverticula, whereas post-resection angiography revealed the "coral reef" vascular anomaly consistent with angiodysplasia. Histology confirmed the presence of both diverticular disease and angiodysplasia. This case report highlights the importance of considering a vascular anomaly in patients presenting with rectal bleeding despite the presence of another radiologically demonstrable anatomical lesion. (J Clin Pathol 1995;48:272–275)

Keywords: Angiodysplasia, diverticular disease, vascular anomaly.

The term angiodysplasia was first used by Athanasoulis et al in 1978 and the condition has since emerged as a distinct clinicopathological entity. Angiodysplasia is recognised on the basis of clinical presentation and endoscopic and angiographic findings. Histological confirmation is made possible by appropriate postoperative barium–gelatine vascular injection.

Angiodysplasia is a frequent vascular anomaly of the large bowel, particularly in the elderly, but only a minority of patients present with prolonged lower intestinal bleeding.

Although angiodysplasia is regarded as a separate vascular entity, its classification and differentiation from other vascular conditions still presents a clinicopathological problem. Much of this relates to our poor understanding of the pathogenesis of angiodysplasia. Several clinical associations have been described. These include aortic stenosis (more precisely the association of rectal bleeding with aortic stenosis), chronic renal failure and cirrhosis. Angiodysplasia is currently thought to represent a degenerative vascular change with the histological hallmark of mucosal vascular ectasia. However, cases have been described in the young, questioning this degenerative pathogenesis.

Here, we present a case in which an unusual form of angiodysplasia occurred in association with congenital right sided diverticular disease. This dual pathology highlights the importance of considering a vascular abnormality of the intestine as a cause of rectal bleeding despite the presence of another radiologically demonstrable anatomical lesion.

Case report

A 74 year old white man presented with a four day history of rectal bleeding, passing stools mixed with fresh, bright red blood. He was otherwise well with no other gastrointestinal symptoms. He had previously had a villous adenoma of the rectum resected endoscopically three years before. On examination, he was haemodynamically stable with a haemoglobin concentration of 13–9 g/dl and a precordial ejection systolic murmur that radiated into the axilla, interpreted as aortic stenosis. There was no evidence of circumsoral or gingival telangiectasia and no family or past medical history of rectal bleeding, telangiectasia, epistaxis, or haemoptysis. There were no signs of congestive cardiac failure, renal insufficiency, portal hypertension, or varices.
Angiodysplasia and caecal diverticulosis

Angiodysplasia is a condition characterized by abnormal vascular patterns in the intestinal wall. It is often associated with the formation of intravascular clots which would preclude adequate visualisation at angiography.

A barium-gelatin mixture (100 ml; 40°C) was injected into the largest supplying artery of the resected specimen until all vessels were filled. Radiographs were then taken (fig 1). The process was then repeated on the venous side. After injection, the bowel was washed out and filled with 10% formal saline until it was inflated to normal size. The opened ends were closed with elastic bands. The injected specimen was pinned to a cork board and fixed in formalin for 24 hours. After fixation, the bowel was opened lengthwise along the mesenteric border as angiodysplastic lesions are usually found on the antimesenteric border opposite the ileocaecal valve. Mucosal lesions were identified by careful examination with a hand lens and comparison with the specimen radiographs. Appropriate areas were then sampled, embedded in paraffin wax, sectioned, and stained with haematoxylin and eosin and Lawson’s Elastic Van Gieson.

**HISTOPATHOLOGICAL FINDINGS**

The right hemicolecction specimen consisted of 24 cm of ascending colon, caecum, normal appendix, and 4 cm of terminal ileum. Macroscopically, multiple diverticula, measuring up to 1 cm in diameter, were present throughout the specimen particularly in the caecum and proximal ascending colon. No abnormal vessels could be seen in the mucosa before or after angiography.

The histological findings confirmed the presence of multiple right sided diverticula. These were “true” diverticula surrounded by attenuated fibres of the muscularis propria with little overall muscular hypertrophy, suggesting a congenital origin. The mucosa showed focal active chronic inflammation with cryptitis and occasional crypt abscesses, all features of a focal acute on chronic diverticulitis. The crypt architecture and goblet cell complement were both well preserved with no sign of inflammatory bowel disease. Numerous widespread and multifocal ecstatic vessels were present in the lamina propria, submucosa, muscularis propria, and the serosal adipose tissue. Ecstatic thin-walled vessels could be seen traversing an intact muscularis mucosae. At least one of these vessels was large and thick walled (fig 2), more in keeping with a congenital arteriovenous malformation than the thin-walled ecstatic veins, venules and capillaries more characteristic of angiodysplasia. However, it is worth emphasising that these vascular lesions were widespread and multifocal and indeed the vast majority of ectatic vessels almost certainly represented dilated pre-existing vasculature, quite unlike those found in arteriovenous anomalies.

Despite some atypical histological features, it was thought that this specimen fitted best into the diagnostic category of angiodysplasia. In addition, a careful search for the clinical features of the telangiectatic syndromes was negative, excluding the Osler–Rendu–Weber syndrome. Some authors have stressed that...

**Figure 1** Radiograph of right hemicolecction specimen after vascular injection with barium-gelatin mixture. The “coral reef” vascular pattern of angiodysplasia is seen throughout the caecum and appendix (× 0.5).

**Methods**

**Vascular imaging and gelatine casting of vascular ectasia in the resected specimen**

Preoperative intestinal angiography was not performed. However, at operation the superior mesenteric artery was washed out with heparin–saline solution to prevent the formation of clots which would preclude adequate visualisation at angiography.

A barium enema, carried out shortly after admission, demonstrated caecal and right sided colonic diverticular disease but no obvious mucosal abnormality. There were no diverticula present distal to the mid-transverse colon. These findings were confirmed at colonoscopy, where no obvious angiodysplastic lesions were identified.

Within one month the patient’s symptoms returned and on re-admission his haemoglobin concentration had dropped to 10.2 g/dl. In view of the previous findings an extended right hemicolecction was performed when diverticula were confirmed from the caecum to the mid-transverse colon. The patient remains well with no further rectal bleeding at six months follow-up.
the variations in the histopathological findings in cases of angiodysplasia may reflect stages in the evolution of the lesion.11

Discussion
Although described under a variety of names, angiodysplasias are telangiectasias and consequently the normal honeycomb microvascular pattern of the mucosa is maintained.12 According to most authors the histological diagnosis of angiodysplasia relies on the presence of mucosal vascular ectasia with associated prominently dilated capillaries penetrating the muscularis mucosae.2412 In this case these criteria are satisfied; however, the presence of large, thick walled vessels in the submucosa is not described in the classic case of angiodysplasia. The appearances, although atypical, still fit best with a diagnosis of angiodysplasia. The differential diagnosis of angiodysplasia includes other varieties of telangiectasias, arteriovascular malformations and haemangiomas (although the latter two would be more localised). Mucosal lymphoangiectasia superficially resembles angiodysplasia but the vessels do not contain blood and there is no associated submucosal venous ectasia. It should also be noted that conditions with give rise to chronic venous congestion—for example, congestive cardiac disease, cirrhosis and portal hypertension, although usually more widespread, may cause diagnostic confusion and should be carefully excluded on clinical grounds.

Other causes of colonic telangiectasia include Osler–Rendu–Weber, Sturge Weber and Bonnet–Dechaume–Blanc syndromes.13 Of these, Osler–Rendu–Weber syndrome is most frequently associated with telangiectasias of the gastrointestinal tract; its distinction from angiodysplasia relies heavily on the other features of the syndrome, namely a family history, childhood epistaxis and telangiectases of the oral mucosa and lips. Unlike angiodysplasia, the vascular abnormalities in this syndrome may implicate the muscularis propria and serosa of the bowel.13 Some authors maintain that the histological feature of a sclerotic perivascular stromal component is characteristic of Osler–Rendu–Weber syndrome and helps to differentiate this from angiodysplasia.1 However, few authors have found this to be specific and to date, it is fair to say that the distinction of angiodysplasia from other colonic telangiectasia is not usually possible on histological grounds alone.4

Arteriovenous anomalies of the intestine tend to have a diffuse infiltrative pattern and the vessels show the features of both arteries and veins. Haemangiomas of the bowel comprise a proliferation of abnormal vascular channels and lack the normal organoid appearance of an angiodysplasia.

Aortic stenosis was identified in this patient and its association with lower intestinal bleeding has been described before.1415 The triad of aortic stenosis, recurrent lower intestinal bleeding and anaemia was first described in 1958 by Heyde.2 It is unclear whether aortic stenosis forms a true association with angiodysplasia or merely represents a predisposing condition to vascular congestion or an acquired bleeding diathesis. Recent observations have demonstrated a link with acquired Von Willebrand’s disease, possibly representing a form of reversible consumption of large clotting factors within the unusual flow dynamics of the ectatic vessels producing a bleeding diathesis.4 Alternatively, the relation may reflect a state of low perfusion pressure and consequent ischaemia with an increased likelihood of angiodysplasias to bleed and present for investigation.2 It has been suggested that the bleeding may be relieved by aortic valve replacement1617 or with medical intervention in the form of beta blockers.

The importance of immediate postoperative heparin flushing and subsequent barium–gelatin angiography of unopened resected specimens is emphasised as these thin-walled vessels may collapse and easily be missed in...
Angiodysplasia and caecal diverticulosis

Pounder et al discussed various techniques to demonstrate vascular lesions. Bearing in mind that some of these conditions may rely on vascular dynamics and clinicopathological syndromes it is imperative that both clinical and pathological information be combined to arrive at a correct diagnosis.

In conclusion, both surgeons and pathologists should be aware of the possibility of a vascular anomaly of the intestine in patients with fresh rectal bleeding. The presence of a radiologically demonstrable anatomical abnormality such as diverticular disease, particularly in present in the right hemicolon.

We would like to thank Mr C Jeal and Mr A Wilson for assistance with photography.


B cell signet-ring cell lymphoma of bone marrow

W G McCluggage, H Bharucha, M El-Agnaf, P G Toner

Abstract

A case of signet-ring cell lymphoma affecting the bone marrow and diagnosed by bone marrow trephine biopsy is reported. Normal marrow was replaced totally by cells with large central vacuoles, many of which displaced the nucleus to the periphery of the cell, imparting a signet-ring appearance. Initially, the favoured morphological diagnosis was metastatic signet-ring adenocarcinoma, but on immunocytochemistry the tumour cells were strongly positive for CD45 (leucocyte common antigen) and the B cell marker CD20 (L26). Electron microscopy revealed electron-lucent vacuoles with no discernable internal structure. The tumour was classified as a high grade centroblastic lymphoma using the upgraded Kiel classification. Immunocytochemistry showed the tumour to be of B cell lineage. The patient subsequently responded to chemotherapy. Here, we report a further case of signet-ring cell lymphoma affecting bone marrow and diagnosed by bone marrow trephine biopsy. Despite treatment with combination chemotherapy, the patient died during an episode of septicaemic shock within two months of presentation.

Case report

A 70 year old man was referred by his general practitioner for investigation of central abdominal pain, nausea, vomiting, night sweats, loss of appetite, and weight loss of approximately one stone.

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In a previous issue 'Talbot et al reported the first case of signet-ring cell lymphoma affecting bone marrow. The tumour presented as multiple lytic lesions in the lumbar sacral spin and was diagnosed by a computed tomography (CT) guided bone biopsy. Morphologically, the tumour was classified as a high grade centroblastic lymphoma using the upgraded Kiel classification. Immunocytochemistry showed the tumour to be of B cell lineage. The patient subsequently responded to chemotherapy. Here, we report a further case of signet-ring cell lymphoma affecting bone marrow and diagnosed by bone marrow trephine biopsy. Despite treatment with combination chemotherapy, the patient died during an episode of septicaemic shock within two months of presentation.

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