Angiolymphoid hyperplasia with eosinophilia in the colon: a novel cause of rectal bleeding

D M Berney, M P Griffiths, C L Brown

Abstract

Angiolymphoid hyperplasia with eosinophilia (epithelioid haemangioma) is an uncommon but distinctive lesion seen principally in the skin. A case of severe gastrointestinal haemorrhage in a 63 year old male is reported, which necessitated a right hemicolectomy. A lobulated nodule was seen macroscopically that had the histological appearance of angiolymphoid hyperplasia with eosinophilia, with sheets of lymphocytes and eosinophils associated with many vessels showing plump and pleomorphic endothelial cells. This is the first reported case of this entity in the large intestine.

(*J Clin Pathol* 1997;50:611–613)

Keywords: angiolymphoid hyperplasia; eosinophilia; colon

Angiolymphoid hyperplasia with eosinophilia (epithelioid haemangioma) is a familiar lesion that presents as a skin lesion usually around the head and neck. There are reports of this entity occurring in the mouth^{1,2} but the lesion has not been reported elsewhere in the gastrointestinal tract. Angiolymphoid hyperplasia is often associated with an underlying vascular malformation. A case is reported illustrating that this reactive inflammatory condition may lead to severe gastrointestinal haemorrhage, and demonstrates that this unusual lesion may occur at unexpected sites.

Case report

A previously well 63 year old African male was admitted as an emergency complaining of rectal bleeding. He had a six hour history of two bouts of profuse bloody motions associated with light headedness. He had no history of previous gastrointestinal problems and there was no significant history of non-steroidal anti-inflammatory use.

On examination the only positive finding of note was mild left sided abdominal discomfort. Rectal examination was normal. Sigmoidoscopy revealed profuse bleeding but no bleeding site was seen. All haematological and biochemical investigations were within normal limits. The patient had two further bouts of rectal bleeding requiring resuscitation with two units of colloid. Colonoscopy determined the source of bleeding as the right side of the colon, although the precise site was not identified. Immediate laparotomy and right hemicolectomy was performed; the patient made an unremarkable recovery.

PROCEDURES

The resected bowel was fixed in buffered formalin. The caecum and ascending colon was 35 cm in length. The terminal ileum was 2 cm in length. Macroscopically a multilobulated nodule 1.6 cm in diameter was noted 15 cm from the distal colonic excision margin. No other abnormality was seen.

Blocks of the nodule and the macroscopically normal bowel were taken and embedded in paraffin. Sections were stained with haematoxylin and eosin, van Gieson, and periodic acid Schiff. The sections were also studied immunohistochemically using the avidin-biotin-peroxidase technique using antibodies against CD34 (Q BEND 10, Bionostics, Bedfordshire, UK), factor VIII (Dako, High Wycombe, UK), and CAM 5.2 (Becton Dickinson, Oxford, UK).

MICROSCOPIC FINDINGS

Microscopically, the nodule comprised inflammatory cells infiltrating the submucosa in a lobular distribution, extending through the muscularis mucosae into the lamina propria and the muscularis propria (figs 1 and 2).

The inflammatory cells principally comprised lymphocytes and eosinophils that surrounded many small capillaries, many of which had a plump "hobnail" endothelial lining (fig 3). Focal ulceration was noted. Immunohistochemistry showed these vessels to be strongly positive for CD34 and factor VIII but negative

Figure 1 Low power view showing the lobular distribution of the inflammatory cell infiltrate and a large artery at the base of the lesion.

Department of
Histopathology and
Morbidity, The
Medical and Dental
School of St
Bartholomew's and the
Royal London
Hospital, The Royal
London Hospital,
London E1 1BB, UK
D M Berney
C L Brown

Department of
Surgery
M P Griffiths

Correspondence to:
Dr Berney.

Accepted for publication
8 April 1997

Figure 2 Dense infiltrate of inflammatory cells in the submucosa with an adjacent distorted artery.

for CAM 5.2. A prominent distorted medium sized artery was seen beneath the main infiltrate. An elastic van Gieson stain showed disruption and reduplication of the internal elastic lamina and intimal hyperplasia of this artery (fig 4). The regional lymph nodes showed non-specific reactive changes.

Sections from the remainder of the colon showed no other abnormality. In particular there was no vascular ectasia or evidence of inflammatory bowel disease.

Discussion

The histological findings in this case are identical to those seen in the skin in angiolymphoid hyperplasia with eosinophilia (epithelioid haemangioma). This entity typically occurs during early to mid adult life and affects more women than men. Most lesions are situated around the head and neck. Bleeding is a common secondary feature. Occasionally there is local lymph

Figure 3 High power view showing sheets of lymphocytes and eosinophils including vessels lined with "hobnail" endothelial cells.

node enlargement and peripheral eosinophilia, although an infectious agent has never been isolated. One third of the lesions recur and one case was reported in which metastasis to a regional lymph node occurred³; however, large series illustrate benign behaviour.⁴ Although some authors consider these lesions to be neoplastic, it has been shown that 60% of cases are associated with a large vessel showing mural damage or rupture.^{5,6} This is identical to the findings in the present case and we believe that these lesions are reactive in nature, possibly arising secondary to damage and repair of an artery or vein. For this reason we do not prefer the alternative name for this lesion, epithelioid haemangioma, which implies a neoplastic entity.

Angiolymphoid hyperplasia with eosinophilia is frequently confused with Kimura's disease with which it has some superficial morphological similarities.⁷ However, deposits of Kimura's disease contain numerous lymphoid follicles and have attenuated endothelial cells lining the vessels⁸ while large distorted vessels are not seen. Kimura's disease is also invariably associated with a peripheral blood eosinophilia. To our knowledge, angiolymphoid hyperplasia with eosinophilia has not been reported in the large bowel. However, cases are reported with lesions within the oral cavity² and specifically on the tongue.¹ Vascular ectasias and angiodysplasia are well recognised in the large bowel as a cause of rectal bleeding and Dieulafoy's vascular malformation has similar large vessels to our case running in the submucosa. However, there are no reports of an associated inflammatory cell infiltrate of this nature in these malformations and Dieulafoy's vascular malformation is seen primarily in the stomach. The alternative diagnoses of inflammatory fibroid polyp and eosinophilic colitis were also considered. While inflammatory fibroid polyps frequently possess a marked infiltrate of eosinophils, they have not been reported as showing the marked arterial changes that were seen at the base of this lesion, nor the prominent endothelial changes. Also, there was no oedema or spindle cell stroma in our case, features that are well described in inflammatory fibroid polyp. The possibility of eosinophilic colitis is ruled out because of the focal nature of the lesion. Histopathological recognition of angiodysplasia is frequently difficult in resected specimens because of the collapse of

Figure 4 Artery at base of lesion showing severe distortion and reduplication of the internal elastic lamina.

vessels. In contrast, this lesion was easily identified by the nodule seen macroscopically. The propensity of angiolymphoid hyperplasia with eosinophilia to bleed was seen in this case, highlighting a potentially fatal outcome of an unusual lesion previously unreported at this site.

- 1 Razquin S, Mayayo E, Citores MA, Alvira R. Angiolymphoid hyperplasia with eosinophilia of the tongue: report of a case and review of the literature. *Hum Pathol* 1991;22:837-9.
- 2 Toeg A, Kermish M, Grishkan A, Temkin D. Histiocytoid hemangioma of the oral cavity: a report of two cases. *J Oral Maxillofac Surg* 1993;51:812-14.
- 3 Reed RJ, Terazakis N. Subcutaneous angiolymphoid hyperplasia with eosinophilia (Kimura's disease). *Cancer* 1972;29:489-97.
- 4 Olsen TJ, Helwig EB. Angiolymphoid hyperplasia with eosinophilia: a clinicopathologic study of 116 patients. *J Am Acad Dermatol* 1985;12:781-96.
- 5 Fetsch JF, Weiss SW. Observations concerning the pathogenesis of epithelioid hemangioma (angiolymphoid hyperplasia). *Modern Pathology* 1991;4:449-55.
- 6 Henry PG, Burnett JW. Angiolymphoid hyperplasia with eosinophilia. *Arch Dermatol* 1978;114:1168-72.
- 7 Kung ITM, Gibson JB, Bannatyne PM. Kimura's disease: a clinicopathological study of 21 cases and its distinction from angiolymphoid hyperplasia with eosinophilia. *Pathology* 1984;16:39-44.
- 8 Googe PB, Harris NL, Mihim MC. Kimura's disease and angiolymphoid hyperplasia with eosinophils. Two distinct histopathological entities. *J Cutan Pathol* 1987;14:263-71.

Mucinous cystadenoma of the appendix with raised serum carcinoembryonic antigen concentration: clinical and pathological features

T Shimizu, M Shimizu, K Kawaguchi, W Yomura, Y Ihara, T Matsumoto

Shimizu Women's
Clinic, Takarazuka,
Japan
T Shimizu
W Yomura

Department of
Pathology, Kawasaki
Medical School,
Kurashiki, Japan
M Shimizu

Division of
Gastroenterology,
Department of
Medicine
T Matsumoto

Department of
Obstetrics and
Gynecology, Tazuke
Kofukai Medical
Research Institute,
Kitano Hospital,
Osaka, Japan
K Kawaguchi

Department of
Obstetrics and
Gynecology, Kobe
City General Hospital,
Kobe, Japan
Y Ihara

Correspondence to:
Dr Shimizu, 2-2-4
Minamiguchi, Takarazuka,
Hyogo, Japan 665.

Accepted for publication
13 May 1997

Abstract

A case of mucinous cystadenoma mimicking ovarian cancer is reported. Serum carcinoembryonic antigen (CEA) concentration was raised, and computed tomography of the abdomen and pelvis demonstrated a long oval shaped cystic mass measuring 9 cm in length on the right anterior side of the uterus. Because of possible right ovarian cancer, laparotomy was performed and the mass was found to be a mucinous cystadenoma of the appendix. This case indicates that mucinous cystadenoma of the appendix may show an unusual presentation including its location as well as the high serum CEA, mimicking ovarian cancer. Therefore, gynaecologists as well as gastroenterologists should consider its possibility as a differential diagnosis of the right adnexal mass in a patient without previous appendectomy.

(*J Clin Pathol* 1997;50:613-614)

Keywords: mucinous cystadenoma; mucocoele; appendix; carcinoembryonic antigen; ovarian cancer

An enlarged appendix with luminal dilatation by mucus has generally been called mucocoele.¹ Higa *et al.*,² in 1973, investigated cases of mucocoele of the appendix and they classified their lesions into three groups: mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. These investigators also recommended avoiding the term mucocoele. However, mucocoele is still used for gross description, rather than histological diagnosis.¹ In addition, the term retention mucocoele, also called simple mucocoele, is still applied to a pathological description of the mucinous

dilatation of the appendiceal lumen resulting from any cause other than epithelial proliferation.^{3,4} On the other hand, when an appendiceal adenoma secretes large amounts of mucus resulting in a clinically palpable cystic lesion, the term cystadenoma of the appendix preferably is used.

We report a case of mucinous cystadenoma of the appendix accompanied by raised serum carcinoembryonic antigen (CEA) concentrations. The case was unique because of the location of the tumour, which led to an initial diagnosis of ovarian cancer.

Case report

A 75 year old Japanese woman (gravida 6, para 6) underwent a medical check up that identified raised serum CEA. Two months later, she visited a women's clinic and was referred to our hospital with a putative diagnosis of right ovarian cancer. Laboratory data including CA125 and CA19-9 were all within normal limits, except for a raised value for CEA (17.7 ng/ml; normal range 0-2.5). While there was no mass palpable on the abdomen, physical examination of the pelvis revealed a sausage shaped mass in the right adnexal region measuring 8 cm in length. There was no lymphadenopathy and cytological examination of the cervix and vagina showed no malignancy. Computed tomography (CT) and ultrasonography of the abdomen and pelvis demonstrated an elliptical cystic mass measuring 9 cm in length on the right anterior side of the uterus (fig 1). Barium enema examination showed no abnormalities within the colorectum.

Laparotomy was performed because of possible right ovarian cancer. During the operation, no ascites was noted. Operative findings