A rare cause of small bowel bleeding

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Reviewer
Simon Anderson

If the diagnosis of gastrointestinal bleeding is unclear from gastroscopy and colonoscopy, investigate with video capsule endoscopy for rarer causes.

Introduction

Gastrointestinal (GI) bleeding is considered a medical emergency requiring hospital admission for urgent diagnosis and management. In the majority of cases, a combination of gastroscopy and colonoscopy will result in diagnosis. However, it is not uncommon for the origin of the bleeding to be in the small bowel. In this case, we report a rare cause of bleeding from the small bowel.

Case

A 57-year-old Caucasian woman presented with lethargy and malaise following an episode of malaena. She was haemodynamically stable. Her haemoglobin was 7.5 g/dL, and the clotting profile was normal. She was transfused with four units of blood. Gastroscopy revealed a 5 mm chronic gastric ulcer, which had no stigmata of recent haemorrhage. Therefore, colonoscopy was also undertaken. This showed two small polyps, which were resected, but no bleeding point was identified. The patient remained stable, so she was discharged home with a follow-up for video capsule endoscopy (VCE).

Her past medical history included primary angiosarcoma of the right breast two years previously, which was treated with right mastectomy, partial lymph node clearance and an adjuvant course of radical radiotherapy. She also had a history of hypertension. A year later, she had presented with a single metastatic skin deposit, which was treated with surgical excision.

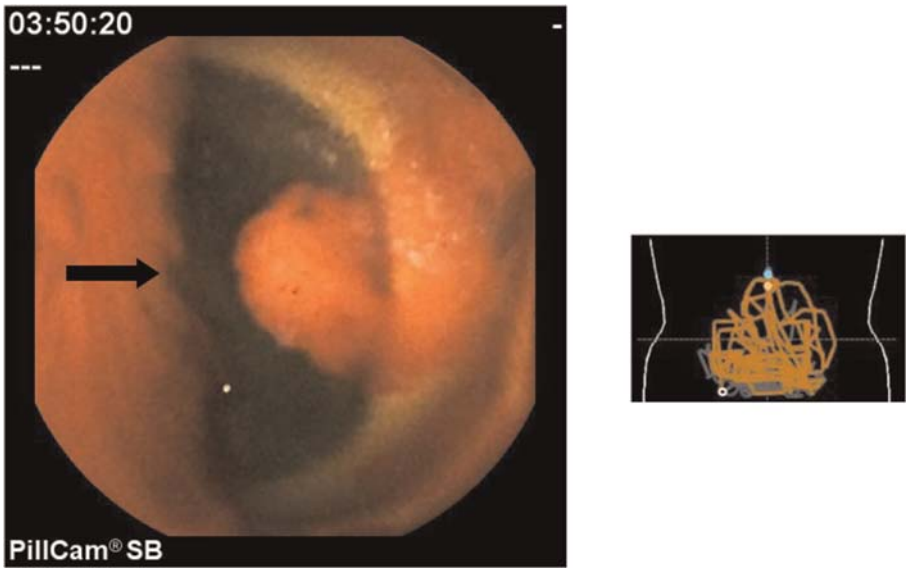
The patient was re-admitted two days later with further malaena. Repeat haemoglobin had dropped to 6.6 g/dL and therefore further blood transfusion was given. An urgent VCE was performed which showed an active bleeding in the small bowel (Figure 1). Computerized tomography angiogram of the abdomen showed multiple metastatic liver deposits, as well as evidence of a tumour at the jejuno-ileal junction of the small bowel (Figures 2a and b). Urgent laparotomy was performed and the bleeding lesion was resected. The patient made an uneventful recovery and was discharged home a few days later. The histological features were consistent with metastatic angiosarcoma (Figures 3a–c).

Four months later, the patient had required further blood transfusion, although she did not report any malaena. She was referred to a specialist Sarcoma unit and commenced on Abraxane chemotherapy, but had progressive metastatic liver disease for which she was treated palliatively with doxorubicin. The patient died of progressive disease, 11 months after the first episode of small bowel haemorrhage, and 35 months after diagnosis of the primary breast tumour.

Discussion

Primary angiosarcoma usually presents with cutaneous deposits, particularly in the head and neck region in elderly patients, or in the limbs of patients with lymphoedema.[1] It is more common in women.[1,2] The majority of intra-abdominal

Figure 1
Image from Video Capsule Study demonstrating a mass lesion in the distal jejunum that was seen to be actively bleeding with overlying blood clot (arrow).



angiosarcomas are found within solid organs such as liver, spleen, adrenals and ovaries, and the disease can be disseminated ('angiosarcomatosis'). There are only a handful of case reports of primary and secondary GI angiosarcomas, most frequently of the small and large intestine, and rarely in the stomach.[2]

Angiosarcoma in the GI tract usually presents with bleeding, anaemia and/or pain, and tends to be at an advanced stage at the time of presentation.[3] Predisposing factors for angiosarcoma include radiotherapy, chronic inflammatory conditions, chronic lymphoedema, long-term dialysis, intra-abdominal foreign body and vinyl chloride, arsenic or thorium dioxide exposure.[1,3] Our patient had no significant medical history prior to the diagnosis of her primary breast angiosarcoma, which appears to represent *de novo* idiopathic disease.

Allison *et al.*[2] described the only case of angiosarcoma metastasizing from a breast primary to the GI tract, within the caecum of a 41-year-old woman. This case shared many similarities with our case including the presentation (GI bleeding and anaemia) and concomitant liver metastases. In this case, treatment included resection of the affected large bowel followed by chemotherapy (Paclitaxel) and radiotherapy. The patient died of disease 33 months after diagnosis.

The histological differential diagnosis of this tumour, particularly with regard to the solid areas with an epithelioid component, includes melanoma, proximal-type epithelioid sarcoma, poorly differentiated carcinoma and epithelioid malignant peripheral nerve sheath tumour, but these tumours can be excluded following immunohistochemical analysis.[2,3] Our case showed the typical architecture of anastomosing vascular channels dissecting collagen, and while there were solid areas focally, these were composed of spindle cells and lacked epithelioid cell morphology.

Treatment of primary and secondary mammary angiosarcoma is primarily with surgery: total mastectomy with or without axillary lymph node dissection, segmental mastectomy or wide local excision.[4] In the cases of high-grade lesions and when metastasis has occurred, chemotherapy is observed to be of some benefit (e.g. anthracycline–infosfamide or gemcitabine–taxane combinations).[4] Radiotherapy has also been administered to patients with breast angiosarcoma, either before resection or after surgery, as in our case.[5]

Figure 2
(a) CT angiogram reconstruction and Figure. (b) CT angiogram (coronal slice of the abdomen). The white arrows illustrating the arterial-enhancing angiosarcoma metastasis arising from the small intestine.

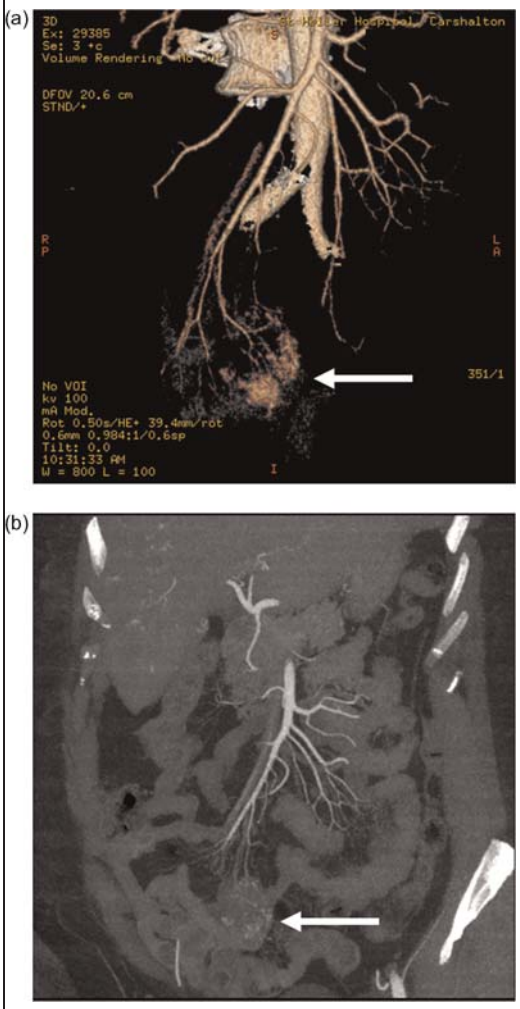
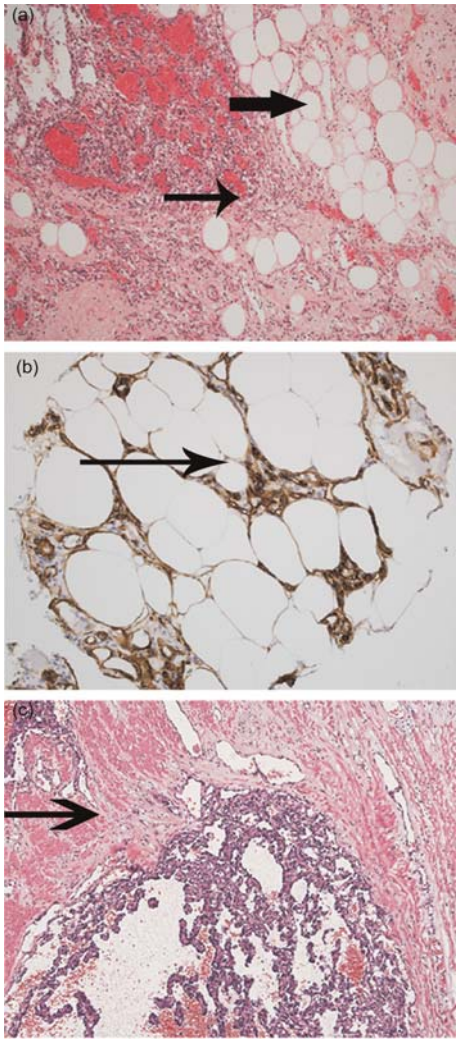


Figure 3
(a) Histology, angiosarcoma, right mastectomy specimen. The primary tumour from the breast is an infiltrative lesion, seen dissecting collagen (thin arrow) and within adipose tissue (thick arrow), and comprising anastomosing vascular channels lined by mildly atypical spindle cells. (b) Immunohistochemistry, angiosarcoma, biopsy from left paraspinal mass. Metastatic tumour, infiltrating paraspinal soft tissue (arrowed), is diffusely positive for CD34, a vascular endothelial marker. (c) Histology, angiosarcoma, small bowel resection. Metastatic tumour is seen infiltrating the muscular is propria (arrowed) of the small bowel. It remains well differentiated, with similar morphology to the primary tumour excised previously from the breast.



Angiosarcomas in the GI tract are generally transfusion-dependent and refractory to conventional therapies. Palliative surgical resection of the haemorrhaging lesions can stop the bleeding; however, complete excision is very difficult due to the infiltrative nature and multicentricity of tumours.[1] Chemotherapy, e.g. taxol and thalidomide, have been used to control bleeding in these GI tumours, but due to the rarity of these malignancies, randomized controlled trials have

not been performed to elucidate the efficacy of these therapies in the GI tract, and so treatment is best planned on a case-by-case basis.

Survival from angiosarcoma may depend on several factors, including tumour size and grade.[4,6] Overall, these tumours behave aggressively and have a high propensity for distant metastasis. Reports of primary and secondary GI angiosarcoma demonstrate a median survival of two months from diagnosis, with uncontrollable haemorrhage the most common cause of death.[2]

In conclusion, primary breast angiosarcomas are rare, and while they show a strong tendency to metastasize, this is the first case documenting spread to the small bowel. In this instance, the metastasis was associated with significant GI bleeding. Selection of appropriate treatment for this patient was a challenge owing to the rarity of these tumours. This demonstrates the need to closely monitor and investigate patients with angiosarcoma, and to be highly suspicious of future metastases, including those occurring at unusual sites.

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