Gastrointestinal bleeding caused by epitheloid sarcoma: A case report

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SUMMARY

Epithelioid sarcoma (ES) of the small bowel is a rare gastrointestinal tumour. We report a case of gastrointestinal bleeding secondary to small bowel ES in a 55-year-old gentleman. After gastroscopy and colonoscopy failed to identify the source of bleeding, we proceeded with computed tomography angiogram of the mesentery, which revealed intraluminal blood clot in the distal jejunum with features of obstruction. This is a rare cause of obscure gastrointestinal bleeding and emphasises the need for additional evaluation in the presence of negative endoscopic findings.

KEY WORDS:
Epithelioid sarcoma, small bowel tumor, obscure gastrointestinal bleeding

INTRODUCTION

Small bowel tumour is very rare, accounting for 1-2% of gastrointestinal neoplasm. Among the sarcomas, the epithelioid variant is exceedingly rare, listed under tumours of uncertain differentiation in the WHO classification of tumour.[1,2] We report a case of small bowel epithelioid sarcoma (ES) which presented with obscure gastrointestinal bleeding, and discuss the role of computed tomography computed tomography (CT) angiography.

CASE REPORT

A 55-year-old Chinese man presented with a four day history of generalised abdominal pain and distension, vomiting and no bowel opening, but was able to pass flatus. He also experienced difficulty in breathing of two weeks duration. His blood pressure was 156/100mmHg, heart rate was 118 bpm, and he had a hemoglobin of 8.0g/dL (normochromic normocytic). Digital rectal examination showed no blood or melena. The abdominal x-ray was unremarkable while the chest x-ray revealed a mass over the right middle lobe, which made us suspicious of metastatic disease. Gastroscopy revealed Forrest 2C ulcers at the lesser curvature and incisura, with bilious fluid in the stomach and duodenum. Colonoscopy findings were grossly normal.

Bilious vomitus ensued and melena appeared on day 2 of admission. Haemoglobin level continued to drop and repeated blood transfusions were given. Repeated gastroscopy done to evaluate the stomach and duodenum revealed no pathology. We then proceeded with CT angiogram of the mesentery. This revealed intraluminal blood clot in the distal jejunum with proximal loop dilatation, which indicated obstruction. However, there was no evidence of active bleeding during the scan (Figure 1).

The patient was subsequently put up for exploratory laparotomy and en-bloc resection was performed. There were multiple intraluminal and extraluminal tumours along the whole length of the small bowel, together with blood clots and multiple enlarged mesenteric and para-aortic nodes (Figure 2). Since the disease was extensive, we only resected the two largest tumours to relieve the obstruction.

The patient's condition rapidly deteriorated over a short period of time and he succumbed to pneumonia and sepsis.

Histologically, the surface of the tumour was ulcerated with a large area of necrosis and lymphovascular invasion. It was surrounded by malignant cells with pleomorphic and large vesicular nuclei, prominent nucleoli and ample of eosinophilic cytoplasm. Immunohistochemical staining demonstrated positive for Vimentin (diffuse), CKAE1/ AE3, EMA and CK7. This supported the diagnosis of epithelioid sarcoma.

DISCUSSION

ES was first described in a paper by Enzinger in 1970. He described this sarcoma as a unique entity and it was frequently confused with a variety of malignant and benign conditions. It has a predilection for distal extremities and is most frequently found in the fingers, hand or wrist.[3] Then in 1997, a more aggressive proximal variant of ES was described by Gillou, which normally present in proximal extremities or axial location of young and middle age adults.[4]

The clinical presentation of ES is usually nonspecific and this remains a challenge in terms of diagnosis and management. Commonly, these patients do not seek early medical attention due to the painless and indolent nature of the tumour, and by the time they present, the tumour may be multifocal.

They may present with slow growing skin lesions or gastrointestinal symptoms that may create surgical emergencies if they go unrecognised, i.e. acute haemorrhage, intestinal obstruction or perforation.[5] ES may dissemination

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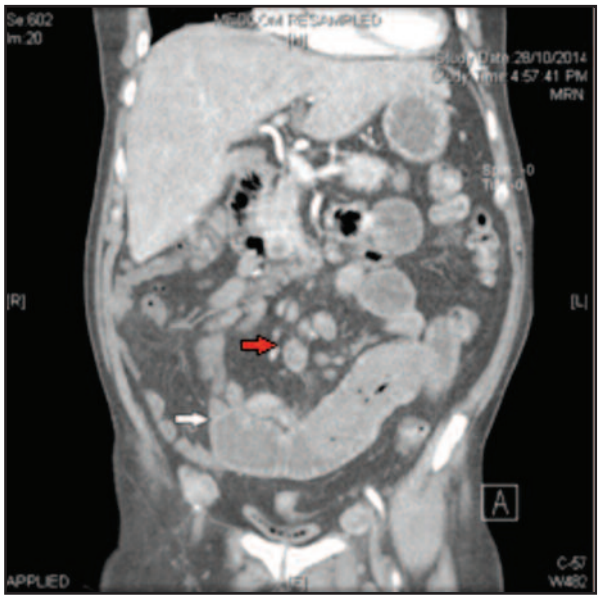

Fig. 1: Sagittal section of CT angiogram of the mesentery showing jejunal loop dilatation with circumferential thickening of bowel wall (white arrow); and multiple enlarged mesenteric and para-aortic nodes (red arrow).

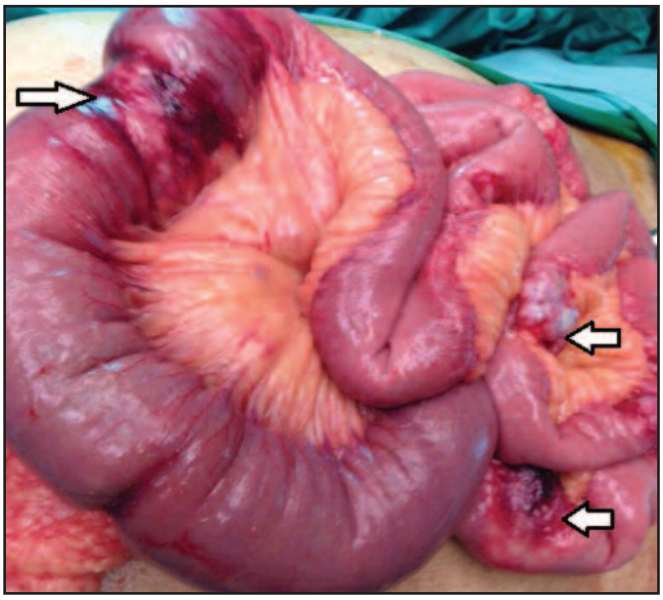

Fig. 2: Multiple extraluminal and intraluminal tumours along small bowel.

via the subdermal lymphatic vessels or the blood stream, and the lungs are the most common site of distant organ metastasis; as seen in our patient where the lung lesion is most likely a metastatic tumour.[3]

Like any other soft tissue neoplasm, diagnosis of ES is made via histopathological examination. However before obtaining the biopsy, initial imaging using CT scan or magnetic resonance imaging (MRI) scan is essential; often to look for the cause of symptoms, to find a tumour or to assess the disease progress. PET scan is also used for its ability to provide functional and structural information, and for identification of unknown primary and unusual metastatic sites. It is not often used for ES but it is seen to be useful in monitoring the response of treatment or progress of the disease. In our patient, CT Angiogram of the mesentery was used to detect the probable cause of the gastrointestinal bleed after negative findings on gastroscopy and colonoscopy.

Due to the rarity of this disease, further study of these lesions has been enhanced in recent years by increasingly available immunohistochemical assays, which allow some differentiation to be established.[5] Chbani *et al* described that vimentin reactivity is present in almost all cases, while pankeratin AE1/AE3 and epithelial membrane antigen were positive in 96% and 98% of the cases respectively.

The prognosis of this disease is very poor due to its aggressive nature, and patients commonly present with metastatic disease.[1] An initial report by Enzinger quoted a local recurrence rate of 85 percent and distant metastatic rate of 30 percent.[3] Palliative surgical resection is one of the consistently proposed treatment options, but this is not the case for everyone.[1] Complete resection is often not possible due to the aggressive infiltrative nature of the tumour, and it has not shown to influence the prognosis of the disease, especially in the case of metastatic disease. There are no sufficiently large trials done to guide us on the management of the disease.

CONCLUSION

A rare cause of small bowel bleeding with nonspecific presentation can be easily mistaken for a benign process. The aggressive nature of ES with nonspecific presentation of the disease may lead to misdiagnosis and improper treatment, thus adversely affecting patient survival, especially when dealing with the small bowel. Therefore, clinicians should bear in mind the role of other imaging modalities in cases of obscure gastrointestinal bleeding, more so if there is a suspicion of malignancy.

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