Vascular tumours of the stomach are rare, representing 0.9%–3.3% of all gastric malignancies.1 Epithelioid haemangioendothelioma (EHE) is one such tumour that can present as upper gastrointestinal bleeding and pose diagnostic difficulties. We report one such case in a 58 year old male patient who presented as an emergency with haematemesis and melaena. In this case report we highlight the rarity of this tumour, the difficulties in diagnosing it preoperatively, and the need for long term follow up in view of the lack of adequate literature regarding management and prognosis. A thorough literature search identified only two reported cases in English language journals1 2 and this seems to be the first case to be recorded in the British medical literature.

CASE REPORT

A 58 year old white man presented as an emergency with a one day history of haematemesis and melaena with no medical history. Abdominal examination was unremarkable and he was haemodynamically unstable. An urgent gastroscopy after active resuscitation showed a 4 mm punched out ulcer along the lesser curvature of the distal body of the stomach. The bleeding was controlled with an injection of 1 ml 1:10 000 adrenaline (epinephrine).

On the following morning the patient had a second episode of significant haematemesis requiring an urgent laparotomy. At operation, an indurated mass was felt in the distal body of the stomach with no other significant abnormality evident in the abdomen. A subtotal gastrectomy with a Roux-en-Y gastrojejunostomy was performed. The postoperative period was uneventful and the patient was discharged on the ninth postoperative day.

Macroscopic examination of the resected specimen showed a 5.0 x 3.5 x 1.5 cm polypoidal lesion on the lesser curve of the stomach (fig 1). Microscopic examination showed a cellular epithelioid neoplasm of the submucosa and muscularis propria lacking apparent mitotic activity (0 per 50 high power fields). Ki 67 staining confirmed a very low proliferation fraction. There was no evidence of epithelial dysplasia in the adjacent mucosa and none of the five lymph nodes retrieved contained metastatic tumour.

The initial histopathological diagnosis was thought to be gastrointestinal stromal tumour (GIST). However, further immunohistochemical studies showed that the tumour cells expressed several vascular markers such as CD31, CD34, and factor VIII related antigen, and CD31 staining highlighted the presence of intracytoplasmic lumina. There was also patchy expression of cytokeratin (fig 2). These findings pointed to

Figure 1  Stomach opened along the greater curve to show the tumour with overlying ulcerated mucosa.

Figure 2  Immunostaining shows CD 34 positivity of many of the tumour cells (background vessels are suitably positive).
Learning points

- This case report serves to highlight the existence of EHE of the stomach as a rare cause of upper gastrointestinal bleeding.
- This tumour can cause diagnostic difficulties preoperatively as biopsies are inadequate because of its submucosal location.
- Immunohistochemistry plays a crucial part in the diagnosis of this pathological entity.
- Neoplasm is generally of low malignant potential with a good prognosis in the absence of histological markers of aggressiveness, however because of definite malignant potential long term follow up is recommended.

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Submitted 29 July 2004
Accepted 3 November 2004

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CONCLUSION

In summary this case report serves to highlight the existence of EHE of the stomach as a rare cause of upper gastrointestinal bleeding. It can cause diagnostic difficulties preoperatively as biopsies are inadequate because of the submucosal location of the tumour. Detailed immunohistochemical evaluation of the full specimen is essential to arrive at a diagnosis. Based on the limited experience of surgical management obtained from review of the literature,1 2 wide excision seems to be adequate for cases with no histological markers of malignant potential (high mitotic figures, spindling of cells, necrosis). The neoplasm is generally of low malignant potential with a good prognosis in the absence of histological markers of aggressiveness, however because of definite malignant potential long term follow up is recommended.
Gastric epithelioid haemangioendothelioma: a rare cause of upper gastrointestinal bleeding

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Postgrad Med J 2005 81: e7
doi: 10.1136/pgmj.2004.027367

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