Multifocal angiosarcoma of the gastrointestinal tract

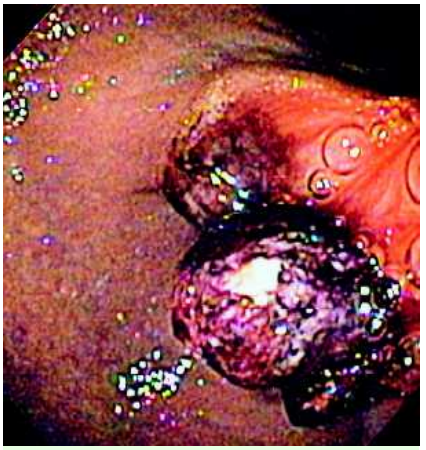

Fig. 1 Esophagogastroduodenoscopy demonstrating an ulcerated hemorrhagic polypoid lesion on the incisura angularis of the stomach.

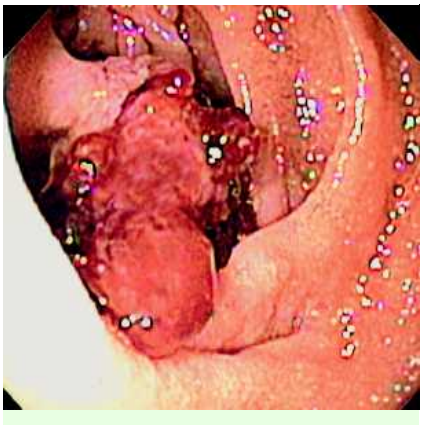

Fig. 2 Esophagogastroduodenoscopy revealing a polypoid lesion in the second portion of the duodenum.

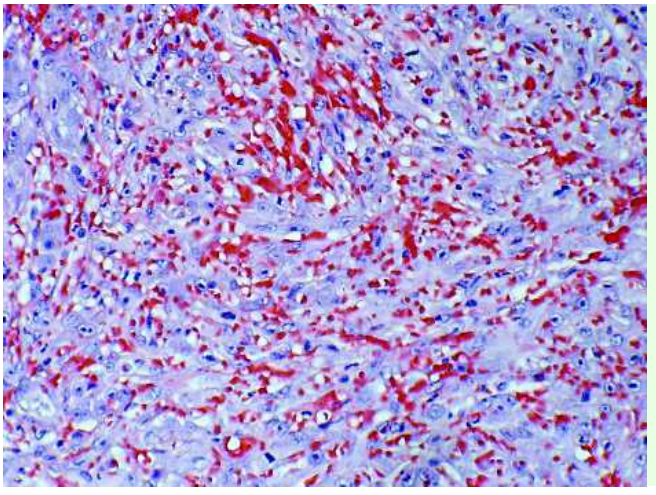

Fig. 3 Duodenum. Photomicrograph showing irregular, anastomosing vascular channels lined by enlarged endothelial cells with epithelioid morphology (hematoxylin & eosin; original magnification × 40).

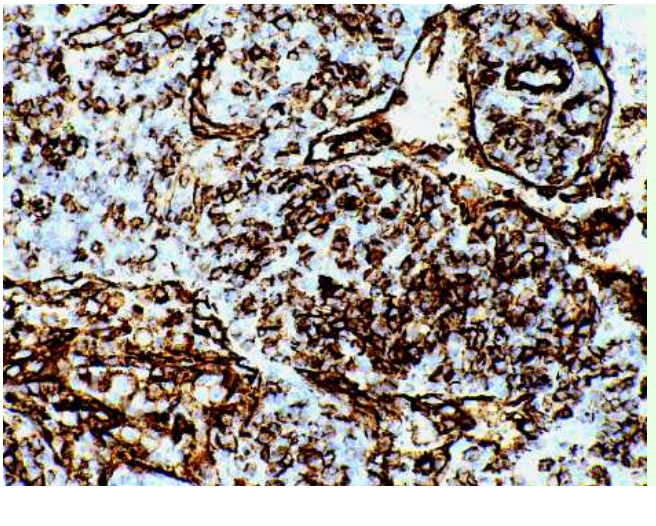

Fig. 4 The neoplastic cells show positive immunoreactivity for CD31 (immunohistochemistry; original magnification × 100).

A 60-year-old African-American woman presented with a history of several weeks of increasing shortness of breath and weakness. On examination, she was hemodynamically stable with heme-positive brown stool. Initial laboratory studies revealed a hemoglobin concentration of 4 g/dL. Upper endoscopy showed active oozing from a 2-cm ulcerated mass on the gastric incisura angularis (**Fig. 1**) and a 1-cm polypoid lesion in the second portion of the duodenum just proximal to the major papilla (**Fig. 2**). Biopsies demonstrated markedly atypical endothelial proliferation suggestive of a high-grade angiosarcoma versus exuberant granulation tissue adjacent to the ulcer (**Fig. 3**). Immunohistochemical stains were positive for CD31 (**Fig. 4**), vimentin, and pancytokeratin, consistent with epithelioid angiosarcoma (EAS). A positron emission tomography scan showed hypermetabolic foci in the stomach and in the region of the duodenum that were highly suggestive of malignancy; no other areas were identified. Due to continued gastrointestinal bleeding and the above findings, the patient underwent an exploratory laparotomy with distal gastrectomy, partial duodenectomy, and gastrojejunostomy. The final surgical pathological study confirmed the diagnosis of EAS.

Primary angiosarcoma of the gastrointestinal tract is extremely rare. These tumors are characterized by an extremely aggressive course, with a high tendency to recur and metastasize, leaving patients with a very poor prognosis [1–4]. Since treatment options for advanced disease are limited, complete surgical resection remains the most crucial factor to achieving a favorable outcome, and this underscores the importance of early and appropriate diagnosis. In patients who present with gastrointestinal bleeding and in whom biopsy reveals a poorly differentiated malignancy, the diagnosis of angiosarcoma should be considered.

Immunohistochemical stains and pathological expertise with gastrointestinal malignancies are often necessary to differentiate angiosarcoma from other malignancies [2,3]. For patients with advanced disease, chemotherapeutic and antiangiogenesis agents are being actively investigated and may have a palliative

role, particularly for patients who become transfusion-dependent [3, 4].

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Bibliography

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