Epithelioid angiosarcoma in the duodenal bulb

Fig. 1 Ulcerated polypoid mass in the duodenal bulb.

Fig. 2 Hemorrhagic lesion in the second duodenal part.

To our knowledge, this is the first case report of epithelioid angiosarcoma (EAS) involving the duodenal bulb.

A 63-year-old male, smoking, saw-mill worker with a medical history of acute myocardial infarction and chronic headache was admitted due to 48 hours of thoracic pain, hemoptysis, and worsening headache. He presented with dyspnea, cutaneous papules on the scalp, chest, right shoulder, and scrotum and 2-cm, firm, painless, subcutaneous nodules on

the forehead as well as some located in the occipital and thoracic regions. Chest and brain computed tomography (CT) scans suggested neoplastic lesions. During the next few days, he developed right lower abdominal pain with a positive psoas sign. Abdominal and pelvic CT scanning demonstrated multiple nodules of the iliacus and piriformis muscles, with histologic findings of tissue necrosis. Following a life-threatening gastrointestinal bleed that was managed with several transfusions, endoscopy revealed a centrally ulcerated polypoid mass in the duodenal bulb, and multiple, slightly raised, umbilicated, lesions not actively bleeding in the second duodenal part (● **Fig. 1** and **2**). Mass biopsies showed neoplastic submucosal proliferation and, in some areas, large epithelioid cells with anastomosing delicate vascular channels. Immunohistochemical staining was positive for endothelial markers (CD31, factor VIII-related antigen) and vimentin but negative for epithelial and lymphoid markers, protein S100, chromogranin, and desmin, indicative of focal EAS. At the end of the diagnostic work-up, adrenal involvement was also found. The patient was administered chemotherapy treatment with ifosfamide and doxorubicin but he died 7 days after the second session.

Primary or metastatic EAS has been very rarely detected with duodenal biopsies [1–4]. Moreover, adrenal and intracranial findings of EAS involving the gastrointestinal tract are extremely uncommon [3,5]. However, the most extraordinary feature in our case is the neoplastic infiltration of the duodenal bulb that revealed the malignancy, supporting the consideration of EAS in the differential diagnosis of neoplasms in the duodenal bulb.

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