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CASE REPORT | SMALL BOWEL

Undifferentiated Carcinoma With Osteoclast-Like Giant Cells: A Rare Cause of Upper Gastrointestinal Bleeding

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ABSTRACT

Undifferentiated osteoclast-like giant cell carcinomas (UOLGCCs) of the digestive tract are very rare, with only a few cases reported in the literature. An 82-year-old man was referred to the emergency department for melena. Endoscopic examination revealed a hemicircumferential ulcerovegetative lesion, involving the bulbar apex and extending to the second portion of the duodenum; biopsies revealed an UOLGCC. The patient underwent transfusion support therapy, and he was proposed for best supportive care. Duodenal UOLGCC is an extremely rare cause of upper gastrointestinal bleeding. Clinical findings and therapeutic approach represent a challenge in this pathology.

KEYWORDS: undifferentiated carcinoma with osteoclast-like giant cells; upper gastrointestinal bleeding; duodenal carcinoma

INTRODUCTION

Extraskeletal undifferentiated osteoclast-like giant cell carcinomas (UOLGCCs) are very rare, with only a few cases reported in the literature of pancreatic, ampullary, and duodenal lesions. Duodenal UOLGCC is an extremely rare cause of upper gastrointestinal bleeding, with only 2 reports in the literature. Given its rarity, the clinical findings and therapeutic approach remain a challenge.

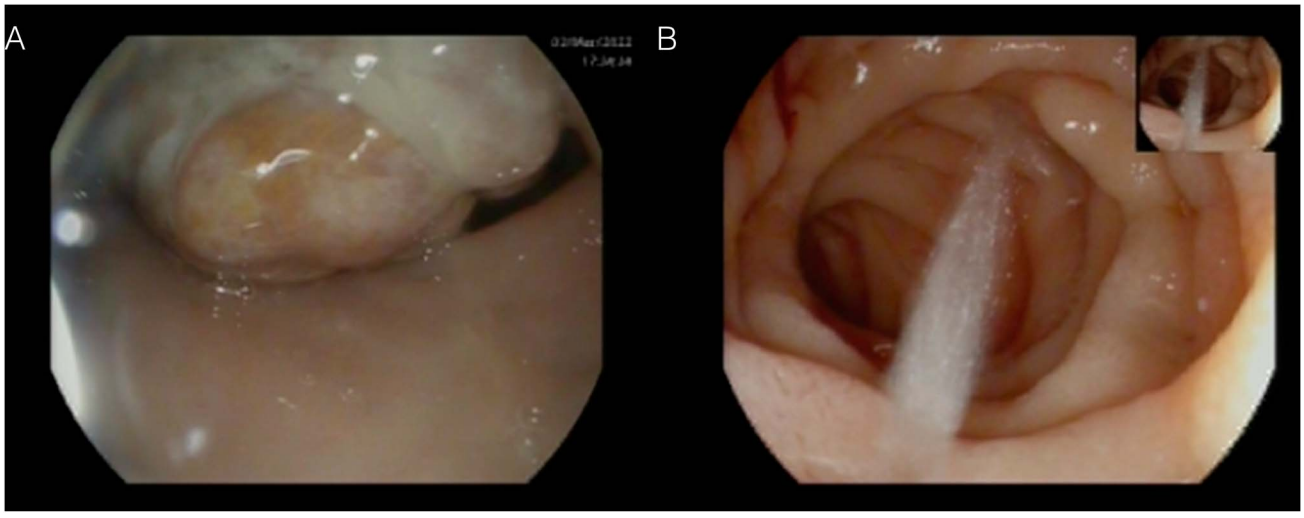

Figure 1. Urgent endoscopy: Hemicircumferential ulcerovegetative lesion is observed, with involvement of the bulbar vertex, leading to a reduction of the luminal caliber (A), but passable with a standard endoscope (B).

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CASE REPORT

An 82-year-old man who was unable to fully perform activities of daily living along with a history of coronary artery disease treated with angioplasty, ischemic stroke, and hypertension presented to the emergency department because of a 3-day history of melena. He was hemodynamically stable with a hemoglobin of 6.1 g/dL.

An urgent endoscopic examination revealed an ulcer-ovegetating, hemicircumferential lesion, with involvement of the bulbar apex and extending to the second duodenal portion, with reduced luminal caliber, but passable with a standard endoscope (Figure 1). Biopsies revealed an undifferentiated carcinoma with multinucleated giant cells of an osteoclast type. Histologically, it was an undifferentiated neoplasm, constituted by epithelioid cells with light-to-moderate pleomorphism, and mixed inflammatory infiltrate, rich in giant cells of the osteoclast type without atypical characteristics and abundant histiocytes (Figure 2).

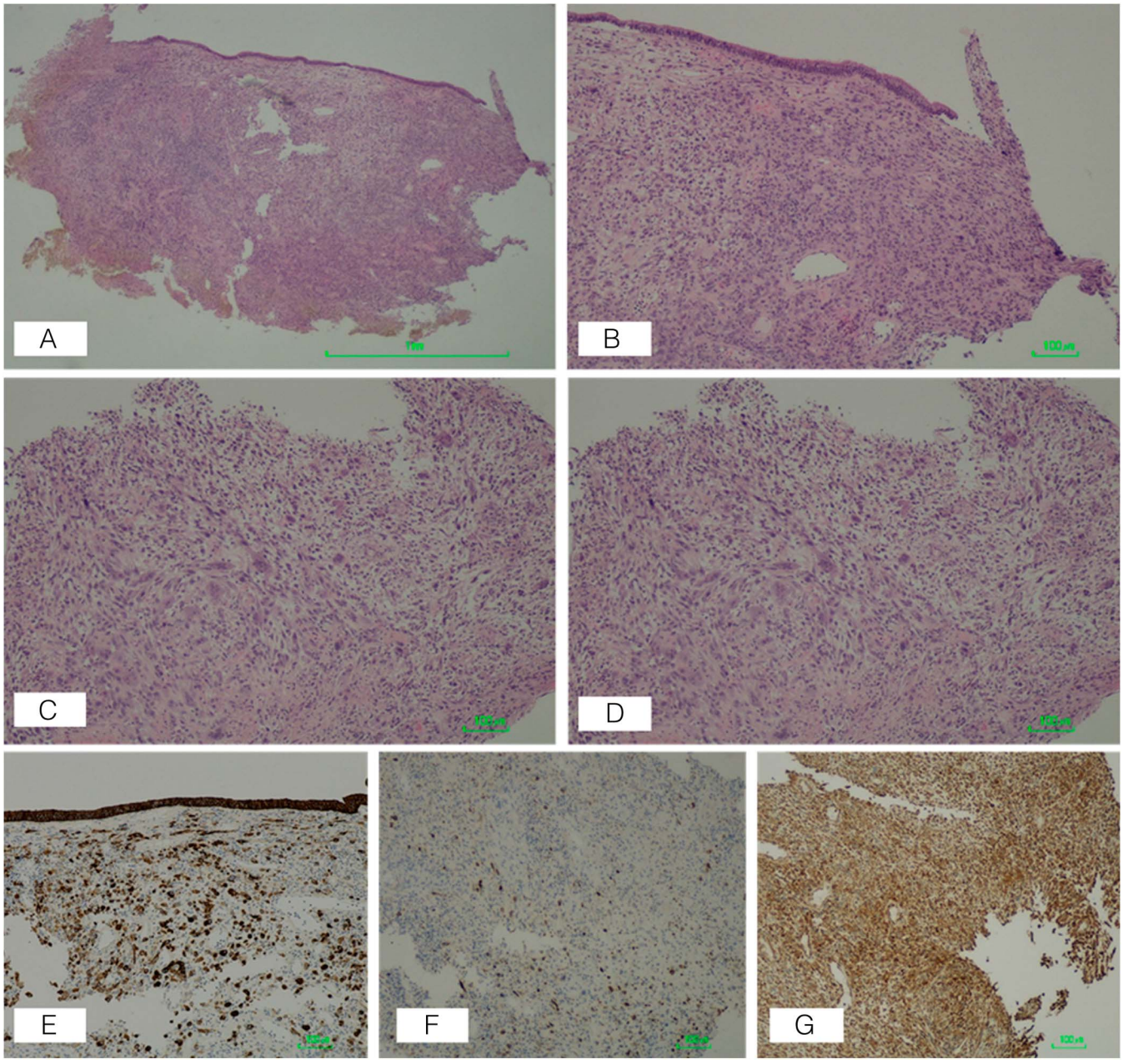

Figure 2. Histological images (stained with hematoxylin-eosin) and immunohistochemistry. Histologically, a neoplasm developed in the lamina propria (A and B), consisting of epithelioid cells with slight-to-moderate pleomorphism (C and D), accompanied by a mixed inflammatory infiltrate, rich in osteoclast-like giant cells (D) and abundant histiocytes (positivity for CD 68); in immunohistochemical analysis, positivity for AE1/AE3 (E) and CK 7 (F) and diffuse positivity for vimentin (G) were observed. The absence of immunoreactivity for the remaining immunomarkers, namely neuroendocrine (synaptophysin and chromogranin A), CK20, CK5/6, Melan-A, pS100, and CD45, was noted.

An abdominal computed axial tomography scan was performed, describing a large tumor formation, measuring at least 9.5 × 6.2 cm, essentially occupying the second portion of the duodenum extending to the third. This lesion presented mainly an endoluminal component, with well-defined contours of the duodenum and no densification of adjacent fat, as well as the absence of involvement of other organs. On the right side, it molded to the hepatic angle of the colon, and on the left side, it presented a close relation with the biliary tract and could also extend to the ampullary region (Figure 3). No lesions compatible with distant metastases were observed.

The patient underwent transfusion support therapy, and the case was discussed by a multidisciplinary team (general surgery, gastroenterology, and oncology). Given the patient's comorbidities and general condition, he was proposed for best supportive care. Currently, he is stable and in follow-up by a palliative care team.

DISCUSSION

Undifferentiated giant cell carcinoma of the duodenal osteoclast type is an extremely rare cause of upper gastrointestinal bleeding, with only 2 reports in the literature to date[1,2] and with no cases reported in Portugal. Although rare, extraskeletal UOLGCCs of the gastrointestinal tract have been reported most in the gallbladder and pancreas.[3]

These tumors are histologically characterized by the presence of a neoplastic epithelial component accompanied by non-neoplastic elements, consisting of osteoclast-like giant cells and histiocytes. In fact, the origin of this tumor is controversial, with some authors advocating a mesenchymal origin[4,5] and others an epithelial origin.[6–8] As previously reported,[9,10] the expression of cytokeratin in the neoplastic component and of vimentin in the poorly differentiated component supports the idea that the origin of the tumor will be mesenchymal; contrarily, the negativity for epithelial markers and the positivity for CD68 in the osteoclast-like giant cells support its histiocytic lineage.

To the best of our knowledge, there are only 2 reported cases of UOLGCC in the duodenal location. The symptoms were subocclusive in one of the cases[2] and of worsening anemia and melena in the other.[1] Indeed, in reported cases of UOLGCC, the neoplasms almost invariably present as large masses,[11] as in the case reported.

Because this is a rare entity, the therapeutic approach is not well established. In patients with surgical conditions, surgery seems to be the choice in cases of UOLGCC of the gastrointestinal tract, especially in the pancreatic location, with variable post-surgical outcomes; there is currently not enough experience to recommend chemotherapy and/or radiotherapy.[12–15]

In the cases of duodenal UOLGCC described in the literature, the therapy was surgical resection, and one of the patients died.[2] Given the rarity of this diagnosis, the prognosis of duodenal UOLGCC is uncertain.

The authors present a case of duodenal UOLGCC, whose clinical presentation was gastrointestinal bleeding (melena); the tumor was a large neoplasm, and given the patient's general condition, best supportive care was decided. The complexity in its histological origin contributes to the difficulty in diagnosis.

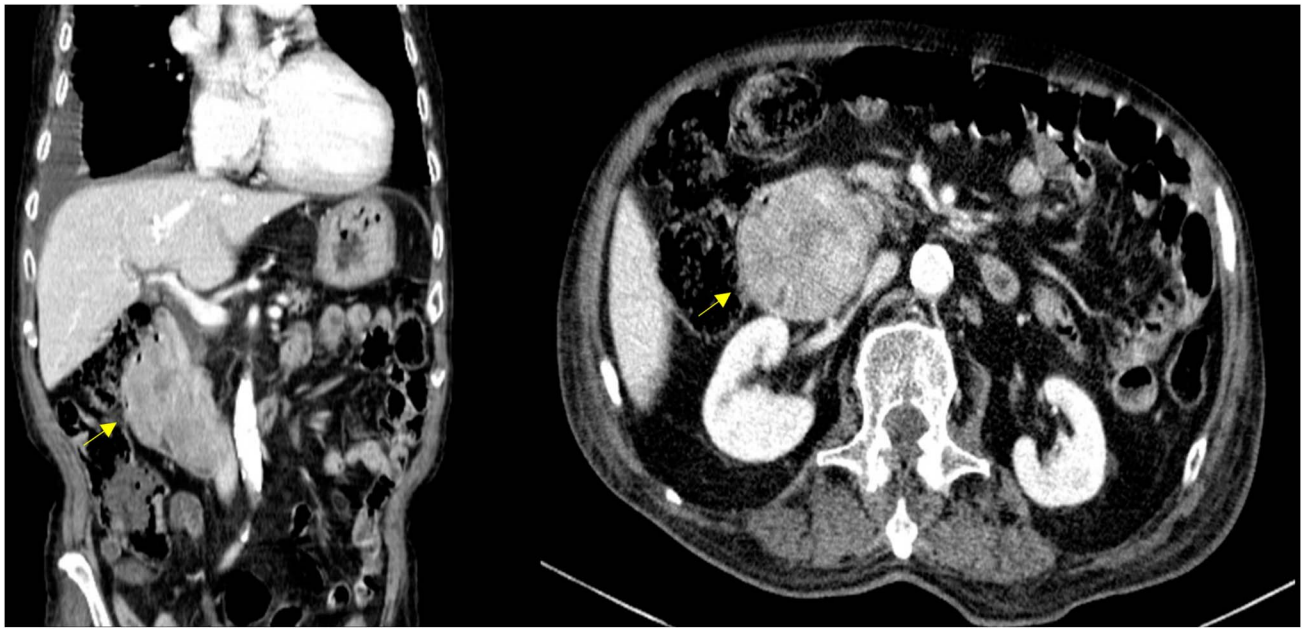

Figure 3. Axial computed tomography revealing a large tumor formation (arrows), measuring at least 9.5 × 6.2 cm, essentially occupying the second portion of the duodenum extending into the third. This lesion presented well-defined contours of the duodenum and no densification of adjacent fat, as well as the absence of involvement of other organs.

Given the exceptionality of these cases, the best therapeutic approach and true prognosis of UOLGCC are currently unknown. Further studies are needed regarding the histological origin of this tumor, so that, with a better knowledge of the tumor, we can perform targeted therapy.

DISCLOSURES

Author contributions: S. Ventura, AC Carvalho, C. Rodrigues, and Â. Domingues: literature research. S. Ventura: manuscript preparation. S. Ventura, P. Rodrigues, and E. Cancela: manuscript editing. E. Cancela, P. Rodrigues, J. Pinho, D. Martins, and A. Silva: manuscript supervising. All authors: manuscript revision/review and final version approval. S. Ventura is the article guarantor.

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