Ménétrier’s disease and Kaposi’s sarcoma in a HIV-positive patient

Dear Editor,

Ménétrier’s disease is an uncommon disease whose etiology remains unknown (1). Its association with Kaposi’s sarcoma (KS) in patients with human immunodeficiency virus (HIV) infection has only been reported in two patients by diSibio et al. (2). Here we report one more case of this exotic association.

Case report

A 39-year-old male presents with burning epigastric pain, nausea, early satiety, vomiting, soft stools, 7 kg weight loss, asthenia, and adynamia for four months. Two weeks before his admission to hospital he had progressive edema in the lower extremities, and an episode of hematemesis. He had a history of syphilis that was treated 18 years ago. Upon admission he had anasarca and mucocutaneous pallor. Examination revealed epigastric tenderness with fluid wave and no masses or visceralomegalies. Purple, vascular-looking, non-bleeding macules were seen on the hard palate, tongue floor, and glans, with laceration of the balano-preputial sulcus and grade-III edema in the lower limbs. ELISA found HIV positivity, which was confirmed with Western Blot, in addition to hypoproteinemia of unclear origin; cardiac, hepatic, renal, and nutritional causes were excluded.

Contrast-enhanced abdominal tomography revealed a poly-poid thickening of gastric walls, and upper digestive endoscopy demonstrated an extensive, diffuse, multinodular, exophytic gastric lesion predominant in the body and fundus. The histopathologic study of the gastric biopsy showed severe foveolar cell proliferation resulting in deep invaginations and replacement of the original oxyntic mucosa. A KS was seen at the lamina propria, which was confirmed by immunolabeling for human herpes virus, type 8 (HHV8) (Fig. 1). Antiretroviral therapy was initiated, and subsequent chemotherapy with anthracyclines was scheduled.

Discussion

Ménétrier’s disease is an idiopathic proliferative disorder of the mucosecretory epithelium in the gastric fundus and body, in association with hypoproteinemia and edema. Mucus hypersecretion leads to nutrient, basic electrolyte, and vitamin malabsorption at the intestinal level, and manifests as a protein-losing gastropathy (3). It is an uncommon condition with only a few hundreds of cases reported in the literature (4), and its joint presentation with HIV infection and KS is even rarer.

To our knowledge, this rare presentation has only been described on two occasions by diSibio et al. (2), who also reported the first association between Ménétrier’s disease and KS. Additionally, DuPrey et al. (5) and Sánchez et al. (6) described two cases in patients with HIV infection and Ménétrier’s disease, positing that HIV-1 might stimulate transforming growth factor α (TGF-α) secretion resulting in continuous activation of the gastric epithelium and increased mucus secretion, hence playing an important role in the pathogenesis of Ménétrier’s disease. However, further studies are needed to confirm such theory.

The findings described herein are relevant because management for the conditions associated with Ménétrier’s disease, in our case infection with HIV and KS, results in regression of the
abnormal mucosa and symptom improvement (7,8). Similarly, Ménétrier’s disease-related endoscopic and histopathologic findings should prompt screening for comorbid KS and/or HIV infection from now on.

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References


Fig. 1. Severe edema of the lower limbs (A). Nodular congestive lesions with hemorrhagic macules in the fundo-corporal gastric mucosa as seen during gastroscopy (B), and polypoid thickening of the gastric wall (C). Severe foveolar hyperplasia associated with Kaposi’s sarcoma in the lamina propria, H&E, 100X (D). HHV8 positivity (immunoperoxidase stain), 400X (E).