A Rare Case of Esophageal Kaposi Sarcoma Causing Dysphagia

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INTRODUCTION

Kaposi sarcoma (KS) is an angioproliferative disorder caused by human herpesvirus-8 in immunocompromised patients. Proliferation and inflammation of the endothelial-derived spindle cells result in a low-grade vascular tumor.

The disease is multifocal, with a course ranging from indolent, with only skin manifestations, to fulminant, with extensive visceral involvement. We are describing a rare case of esophageal Kaposi sarcoma causing dysphagia.

CASE DESCRIPTION

A 57-year-old bisexual male with a medical history of hypertension and a recent diagnosis of HIV not on any medication presented with hypotension, tachycardia, nausea, vomiting, and dysphagia to solids.

On examination, he was noted to have oropharyngeal lesions, and a biopsy of the lesions was obtained. An EGD performed for evaluation of dysphagia (denied odynophagia) showed circumferential erythema, purplish plaques, and friability of the esophagus. Biopsies were obtained from the esophagus. Pathology obtained from the tongue, gingiva, and proximal and distal esophagus returned positive for Kaposi sarcoma with positive staining for HHV-8 and CD31. CD4 count was 31/uL and HSV8 PCR showed 1,000,000 copies. The patient was treated with Bictegravir, emtricitabine, and tenofovir alafenamide therapy

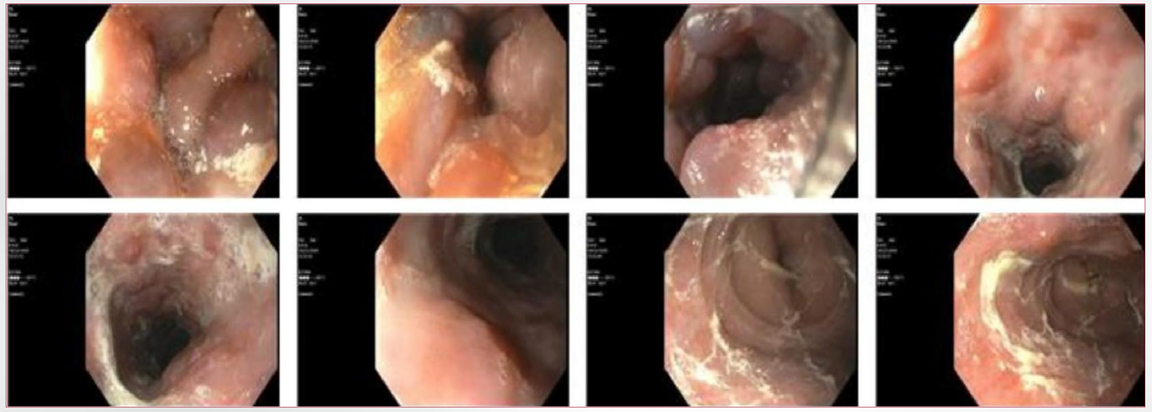

Figure 1: EGD displaying circumferential erythema, purplish plaques and friability of esophagus

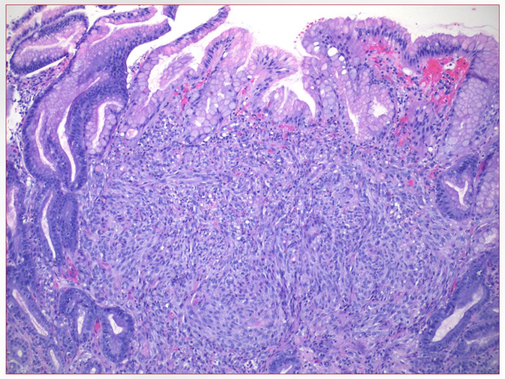

Figure 2: Kaposi sarcoma composed of a spindle cell proliferation within the lamina propria showing myoid features and focal slit-like vascular spaces; foveolar epithelium of the stomach is seen overlying the lesion

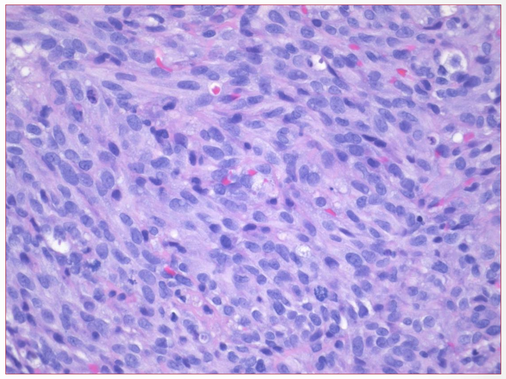

Figure 3: Kaposi sarcoma showing scattered extravasated red blood cells (center and upper right) and mitotic figures

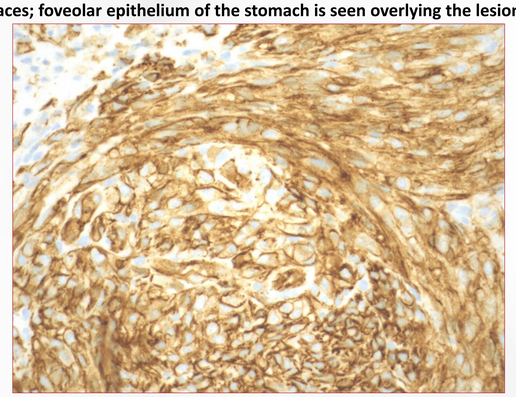

Figure 4: Kaposi sarcoma on CD31 immunohistochemical stain highlights the vascular nature of the lesion

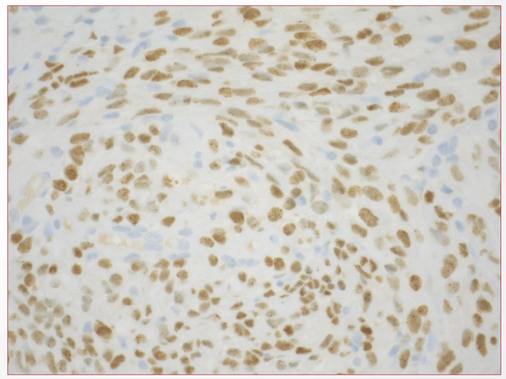

Figure 5: Kaposi sarcoma showing immunoreactivity on HHV-8 immunohistochemical stain

Hematology was consulted, and the patient was started on liposomal doxorubicin. He developed Ventricular fibrillation and cardiac arrest. He was revived, intubated, and moved to the ICU. The patient improved initially but developed aspiration pneumonia. Infectious disease treated him with empirical antibiotics and antivirals.

The patient developed neutropenic fever, Castleman's disease, respiratory failure, acute kidney failure requiring CRRT, and altered mental status. Despite aggressive measures, the patient did not survive.

DISCUSSION

AIDS-related KS varies in its clinical progression and occurs in about 20% of patients with AIDS. The CD4 count will typically be less than 150 cells per cubic millimeter with a high viral load ranging from greater than 10,000 copies per millimeter.

Gastrointestinal manifestation is the most common extra-cutaneous site of KS in AIDS-related cases though rarely does it occur without cutaneous involvement. The majority of the patients are asymptomatic, roughly 75%. Symptomatic patients present with nonspecific findings such as abdominal discomfort, cramps, nausea, vomiting, diarrhea, and upper or lower GI bleeding. KS lesions causing dysphagia are extremely rare. The mainstay of therapy is combined antiretroviral therapy (ART). Local intralesional chemotherapy can be utilized to manage limited lesions. Systemic therapy with liposomal anthracyclines is recommended for patients with advanced or rapidly progressive disease.