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Letter

External ear nodule revealing a disseminated Kaposi disease

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Abstract

Kaposi disease (KD) is an angiogenetic tumor process, characterized by its various clinical aspects. Its pathogenesis is multifactorial and it was attributed to HHV-8 infection. We report an original case of disseminated KD revealed by solitary lesion of external ear in a patient treated by corticosteroids for bullous pemphigoid.

Introduction

Kaposi disease (KD) is the malignant proliferation of endothelial cells of vessels. Its pathogenesis is multifactorial combining genetic, environmental and immune factors. This type of neoplasia has been attributed to human herpes virus Type 8 (HHV-8) infection. It can be classified into four distinct forms: classic, endemic, iatrogenic and AIDS-associated. We report a case of disseminated KD revealed by solitary lesion of external ear in patient with bullous pemphigoid (BP).

Case report

A 64-year-old woman presented with a nodule on the pinna 2 months after being put under corticosteroids (1 mg/kg/day for 6 weeks then decreased) for BP. This nodule gradually increased in size and was associated with abdominal discomfort and general weakness. Clinical exam showed a painless, soft, purple erythematous nodular lesion of the left pinna (Figure 1) and a 3mm purplish blotch lesion on the palate.

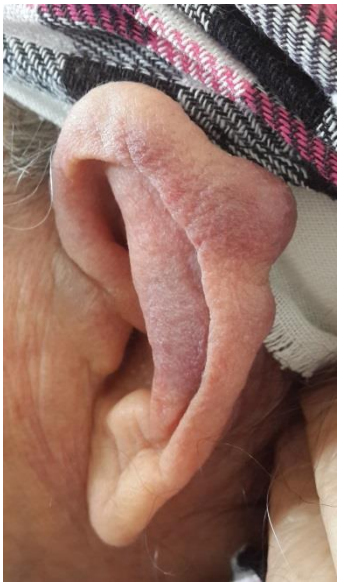

Figure 1. Nodule on pinna

Histological exam showing tumor proliferation with spindle cells and slit-like vascular spaces with red blood cell extravasation (Figure 2). Immunohistochemistry was positive for HHV-8.

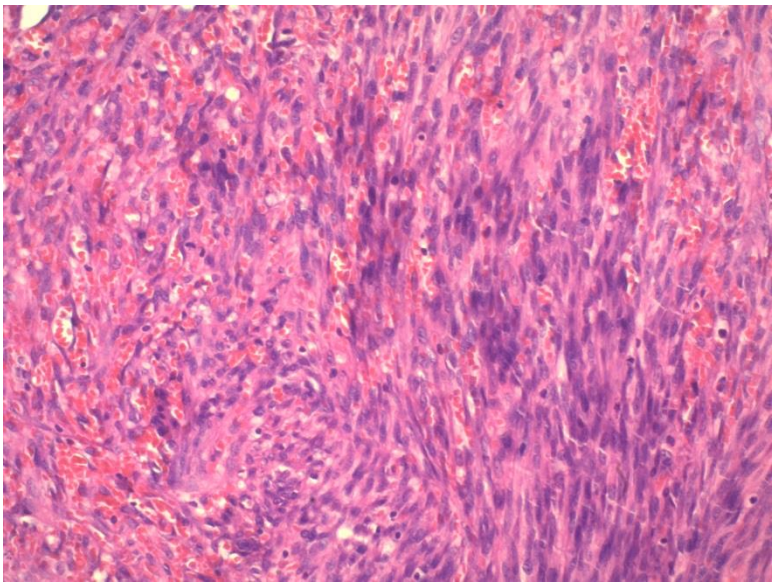

Figure 2. Histology showing tumor proliferation with spindle cells and slit-like vascular spaces with red blood cell extravasation, suggestive of KD (H&E staining, original magnification ×40).

These findings were consistent with the diagnosis of KD. HIV serology was negative. The esophagogastroduodenoscopy showed purple fundic lesions, which upon histological analysis was suggestive of KD.

Given the age of the patient, the context of bullous pemphigoid, asthenia and chronic feeling of intestinal discomfort, associated neoplasia was suspected, hence achieving a colonoscopy and a thoraco-abdominopelvic CT scan. Colonoscopy revealed colonic lesions similar to that observed in the stomach and histological examination of these lesions also suggested KD. Thoraco-abdominal-pelvic CT scan revealed hypodense lesions involving the left lobe of the liver; the larger was 18mm localised at the third segment. A hypodense 10mm lesion of the spleen (Figure 3 a,b) with ring enhancement was apparent with contrast agent.

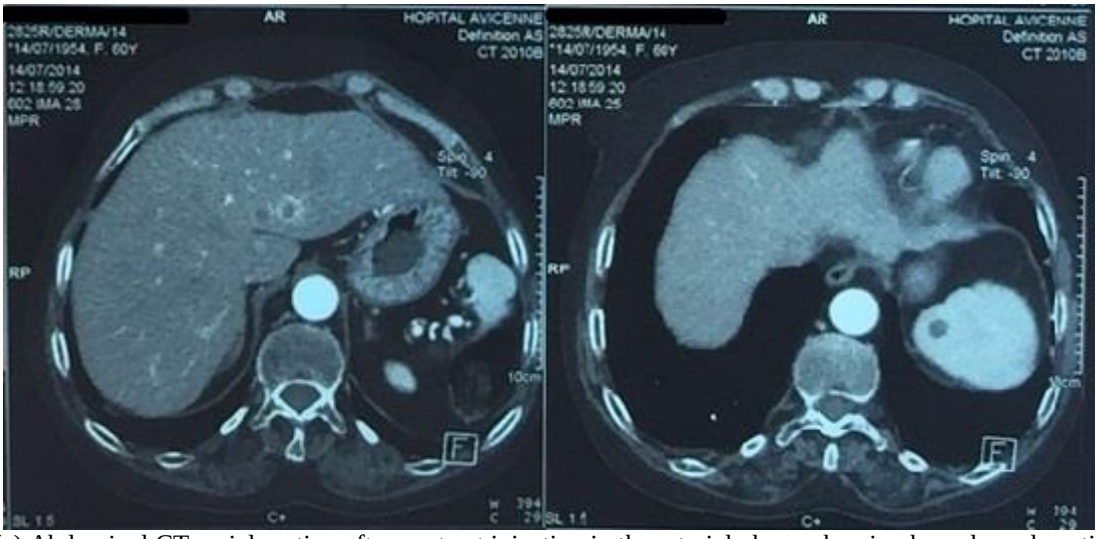

Figure 3 a,b. (a) Abdominal CT: axial section after contrast injection in the arterial phase, showing hypodense hepatic lesions of the segment III with ring enhancement. (b) Abdominal CT: axial section after contrast injection in the arterial phase, showing a hypodense splenic lesions

The CT-guided biopsy of hepatic lesions was in favor of hepatic location of KD. The diagnosis of disseminated KD that affected skin, stomach, colon, liver and spleen was retained. Our patient received Bleomycin every 2 weeks for 3 days (15mg / day), a total of 5 times. Corticosteroid was gradual decreased. We observed an improvement of visceral and skin Kaposi's.

Discussion

Kaposi disease (KD) is an angiogenetic tumor process, characterized by its various clinical aspects. The involvement of the external ear during KD is very rare with only five cases reported in the literature [1-3]. The impact of this location has increased in recent years, probably because of the increased use of immunosuppressive therapy [4].

The case we describe was particularly interesting because this unusual skin location revealed a disseminated KD in a patient with BP. Only 12 cases of KD in patients with BP were reported in PubMed [5]. KD associated with autoimmune bullous dermatosis is regarded as iatrogenic because it is related to the administration of corticosteroids and/or immunosuppressive therapy. The clinical features of iatrogenic KD are similar to AIDS-associated KD with a marked tendency for visceral involvement [6].

The prevalence of visceral lesions during KD is often underestimated as they are usually asymptomatic and are typically post-mortem findings. The gastrointestinal tract is the most highly localized region of extracutaneous KD. Gastrointestinal involvement is usually asymptomatic [4,7], however, with evolution, various symptoms may occur, including abdominal pain, nausea, vomiting, intermittent diarrhea or malabsorption. In rare cases, a large tumor may lead to mechanical obstruction or intestinal perforation [8]. Liver damage, meanwhile, is less common [9]. It usually manifests itself as a tumor that sits in the hilum and diffuses in the liver following vascular routes. Exceptionally it may appear as an intrahepatic tumor or diffuse infiltration of the liver parenchyma [10].

Conclusion

In patients treated with corticosteroids, a single skin lesion may be the unique clinical manifestation of a multifocal KD whish requiring systemic chemotherapy and regular monitoring.

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