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ORIGINAL ARTICLE

Cytopathologic diagnosis of Kaposi sarcoma in unusual clinical settings

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Fine-needle aspiration

Introduction Kaposi sarcoma (KS) is a rare disease that presents as 1 of 4 distinct clinicopathologic subtypes; however, it may present in populations outside those normally encountered. In such cases, it will be important to consider KS in the differential diagnosis, as it may mimic other neoplastic and non-neoplastic entities.

Materials and methods We describe 2 cases of KS, 1 in a patient not clinically fitting any of the 4 subtypes and the other in a patient with atypical presentation in human immunodeficiency virus (HIV)-associated disease. The first is an 81-year-old African American (AA) woman with a history of KS of the leg, who presented with groin lymphadenopathy and the second is a 42-year-old AA man with a known history of HIV infection, no skin lesions, and new axillary lymphadenopathy.

Results Fine-needle aspiration of the groin and axillary lymph node, respectively, showed atypical spindle cells in a lymphoplasmacytic background. The spindle cells were positive for human herpesvirus-8 on the cell block and subsequent lymph node excision. In patients with HIV infection, in addition to reactive and lymphoproliferative processes, KS should be considered. In the former case, the demographic of an elderly AA woman without immunosuppression would not cause concern for systemic KS, but for a metastatic tumor or lymphoma.

Conclusions Cytology is a helpful tool in narrowing the differential diagnosis for spindle cell lesions. With a diagnosis of KS, clinicians would be able to query the clinical history for a possible etiology, such as HIV, and exclude the possibility of metastatic disease.

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Introduction

Before the emergence of the acquired immune deficiency syndrome (AIDS) epidemic, the incidence of Kaposi sarcoma (KS) was limited to a handful of geographical areas and its presentation in the United States was infrequent. Since the early 1980s, the incidence of KS has increased, with most cases involving patients with AIDS, particularly in the demographic of homosexual or bisexual men.[1] However, with the advent of highly active antiretroviral therapy (HAART), the incidence has decreased. The distinction between classical KS and AIDS-related KS is important, as the latter is more aggressive and often involves the viscera.[2]

The histologic features of KS recapitulate those of an angioformative lesion. Namely, there is a proliferation of variably atypical spindle cells, with extravasation of red blood cells and formation of irregular thin-walled vascular channels. Periodic acid-Schiff-positive hyaline globules are often present, but are not pathognomonic of the disease. Another helpful feature is the presence of some irregular channels within pre-existing blood vessels, the so-called promontory sign. The histopathology differs depending on the clinical stage of the disease. The earliest lesions in the patch stage show mild proliferation of vessels in the dermis. Later, as the lesion progresses to the plaque and nodular stages, there is increased proliferation of spindle cells and a lymphoplasmacytic inflammatory infiltrate.

Fine-needle aspiration (FNA) cytology can be useful in the diagnosis of KS in patients with lymphadenopathy, particularly when the pretest probability is high, as in patients with established AIDS or history of human immunodeficiency virus (HIV) infection.[3] Rarely, it can be the first manifestation of a previously unknown HIV infection.[4] However, KS in nonendemic and nonepidemic settings is uncommon, and the diagnosis of KS in patients with lymphadenopathy would not enter the clinician's or cytopathologist's differential diagnosis in the evaluation of an FNA biopsy. In this study, we describe 2 patients with unusual clinical presentations not characteristic of KS. The first is an elderly African American woman with no history of immunodeficiency and the second in a patient with established HIV infection but with an atypical clinical presentation. We present the cases with a discussion of relevant differential diagnoses and highlight pertinent morphologic features and ancillary studies needed to arrive at the diagnosis of KS.

Materials and methods

Smears from both cases were air-dried or alcohol-fixed and stained in the usual fashion with Diff-Quik or Papanicolaou stain, respectively. The residual tissue obtained from needles was rinsed into growth media (Roswell Park Memorial Institute) solution and submitted for flow cytometry.

Following routine hematoxylin and eosin–stained histologic review, immunohistochemical analysis was performed on 4-mm-thick paraffin tissue sections. The following primary antibodies were used: HHV-8, D240, and CD34.

Results

Case 1: The patient is an 81-year-old African American woman with no significant past medical history who presented at our institution with a patch on her lower leg. She underwent a punch biopsy of the skin lesion, which was diagnosed as KS in the nodular stage. Typical histology was present, and many of the neoplastic cells stained for human herpesvirus-8 (HHV-8) immunostain as well as D240 and CD34, supporting the diagnosis. Serum HIV antibody testing was negative on 2 occasions at this time. The patient underwent intermittent chemotherapy and suffered from a recurrence 3 years later. Her disease stabilized after further chemotherapy. She presented 6 years after the initial diagnosis with diffuse lymphadenopathy. FNA was performed on an enlarged right groin lymph node and showed atypical spindle cells with hyperchromatic, variably irregular nuclei and dense, elongated cytoplasm (Fig. 1). The spindle cells were, in areas, arranged in groups, and resembled granulomas (Fig. 2), but were often found singly. There was a background of lymphocytes and plasma cells. These features were suggestive of KS, and an excisional biopsy of the lymph node was recommended for definitive workup. Repeat serum HIV testing was again negative. The lymph node was received by the Department of Hematopathology, and a lymphoma protocol was undertaken to rule out a lymphoproliferative disorder. The protocol involves the examination of touch preparations of the tissue, followed by triage of the specimen for submission of tissue sections for morphology, flow cytometry, and/or cytogenetic analysis. Histology showed extensive replacement of the normal lymph node architecture by proliferating spindled to plump

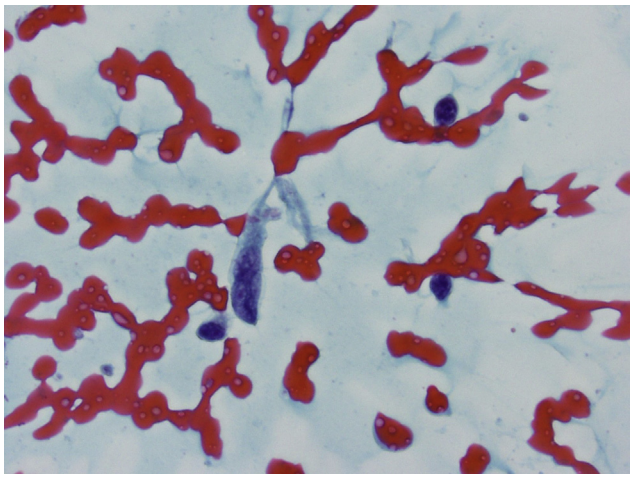

Figure 1 Fine-needle aspiration of the groin lymph node showed spindle cells, some of which had atypical features such as hyperchromasia and nuclear irregularity. (Papanicolaou, 400×)