Kaposi Sarcoma as Initial Presentation of HIV Infection

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Abstract

Context: Kaposi's sarcoma (KS), a vascular tumor that manifests as nodular lesions on the skin and to a lesser extent, the visceral organs, is the most common neoplasm encountered in human immunodeficiency virus (HIV)-infected patients. It consists of an angiosarcomatous change of not only the epithelial and mucous membrane-associated connective tissue in various sites, for example, skin, gastrointestinal system, lungs, and so on, but may also involve non-epithelial organs, such as lymph nodes. Surgical excision is the line of management for the tumor. Case Report: We present one case of a 65-year-old heterosexual Indian male, clinically unsuspected for acquired immunodeficiency syndrome (AIDS) who presented with multiple non-blanching, bluish-red nodules on all extremities, chest, back and bilateral submandibular and cervical lymphadenopathy. Fine needle aspiration cytology (FNAC) was performed from subcutaneous nodule and lymph node. Smears showed hypercellular plump spindle cell groups in a hemorrhagic background. Diagnosis was given as low-grade spindle cell neoplasm consistent with KS, which was later confirmed on histopathology. Conclusion: The first line diagnostic aid of FNAC has several advantages over the traditional biopsy in testing such vascular tumors. The latter is generally needed for confirmation of KS. However, FNAC of such vascular tumors has advantages of better patient compliance, ease of procedure, no recurrences, and safety in immuno-compromised patients. Ancillary studies can be done on aspirates along with polymerase chain reaction (PCR) amplification techniques in confirming the detection of associated human herpes virus-8 (HHV-8) infection with KS.

Keywords: FNAC, HHV-8 virus, Kaposi's sarcoma

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Introduction

Kaposi's sarcoma (KS) is a systemic disease which involves neoplastic cutaneous lesion with or without internal involvement. It was first described by Moritz Kaposi a Hungarian dermatologist. India has high prevalence of human immunodeficiency virus/acquired immunodeficiency syndrome (HIV/AIDS) associated with increased number of KS affecting skin and mucous membranes. Even though KS in HIV/AIDS cases have been reported,[1,2] only very few publications present cytological diagnosis. In this article, we describe an unusual cytopathological presentation of KS in multiple swellings over the trunk, back and submandibular lymph nodes in fine needle aspiration cytology (FNAC) samples, as an initial presentation of HIV infection in a 68-year-old man.

Case Report

A 68-year-old married male presented with multiple swelling on the trunk, back, upper and lower extremities since one year. Lesions started on the trunk and gradually spread to involve upper and lower limbs. Initially lesions were small, flat and then gradually increased in size to become nodular. He also had bilateral submandibular swelling and cervical lymphadenopathy. Patient was heterosexual and moderately built. No history of fever, cough, diarrhoea, weight loss or history suggestive of AIDS sequelae was noted.
On local examination, the swellings were multiple erythematous papules to nodular which were bluish in colour on the back, trunk and both extremities [Figure 1]. The non-blanchable swellings were ranging from 0.5 cm to 7 cm in diameter and firm in consistency. Pallor was present with no hepato-splenomegaly seen and no other systemic findings. The two clinical differential diagnoses were of lipomatosiis and neuromatosis, respectively.

Patient was advised HIV-enzyme-linked immunosorbent assay (ELISA) and he was diagnosed for the first time as reactive to the HIV antibody ELISA test. All routine laboratory investigations were performed along with CD4 count of 239 cells/µl, which was low in this patient. The prothrombin time levels - 12 secs (control 35 secs), activated partial thromboplastin time kaolin levels more than two mins (control 35 secs), serum creatinine - 2.6 mg/dl (0.8 to 1.2 mg/dl), serum glutamic-oxaloacetic acid transaminase levels -130 iu/litre (8-33 iu/litre), serum glutamic-pyruvic transaminase levels -134 IU/litre (4-36 IU/litre). The patient’s ultrasonography (USG) abdomen showed features suggestive of fatty liver with multiple hyperechoic lesions in liver and spleen (suggestive of metastases). Many subcutaneous lymph nodes were also noted on anterior abdominal wall.

The patient was then referred to cytology OPD for FNAC. The FNAC was performed with 22-gauge needle from multiple sites of swellings over the trunk, back and submandibular lymph nodes. The aspirate obtained was hemorrhagic and the cytology smears were hypercellular which showed large cohesive clumps of oval to spindle cells [Figure 2]. The cells had moderate amount of ill-defined, eosinophilic cytoplasm having oval to spindle nuclei with finely granular chromatin, inconspicuous nucleoli and mild pleomorphism. Few mitotic figures were seen. The background showed hemorrhage and macrophages [Figure 3]. Cytological features were suggestive of low-grade spindle cell neoplasm consistent with KS.

The biopsy from the subcutaneous nodule was performed with tumor mass comprised of proliferating blood vessels arranged in slit-like spaces and fascicles. These spaces were lined by endothelial lining separated by spindled cells having moderate amount of eosinophilic cytoplasm and filled with red blood cells with extravasation in surrounding collagen bundles. Perivascular spaces show plenty of lymphocytes, hemosiderin-laden macrophages and neutrophils [Figure 4]. Diagnosis was confirmed on biopsy as KS. Special stain and immunohistochemistry were not performed as patient had financial constraints.

**Discussion**

The KS is an angioformative lesion common in the Mediterranean and Africa. Nowadays, its incidence is increasing due to its association with HIV infections. In India, the incidence of HIV is increasing but prevalence of KS is low as comparative to developed countries may be due to the low prevalence of human herpes virus-8 (HHV-8) in our country.[3] The first case of AIDS associated KS from India was reported in 1993 in a 35-year-old female sex worker. She had multiple painless non-pruritic nodules of varying colours on the right leg with swelling of leg. Since then, a very few cases had been reported amongst Indian patients. Initial reports described KS in homosexual men with AIDS, but many publications have also reported its occurrence in heterosexual males.[4,5] The present case report is of KS in a HIV-positive heterosexual male.

In AIDS related KS has very aggressive clinical course with frequent involvement of lymph nodes, lungs and gastrointestinal tract in 50% of the patients. Involvement
of the lung occurs in 20% of the patients and is the most life-threatening form of the disease.[6]

The causative agent of KS is HHV-8. Its co-infection with HIV promotes the oncogenic capabilities of HHV-8, leading to the development of KS.[7] HHV-8 infects a wide variety of cells, including lymphatic cells and vascular endothelial cells, resulting in the production of lymphangiogenic growth factors. The KS has the ability to develop into lesions of varying morphologic appearance. Lymph node involvement associated with abnormal lymphatics observed in lymphangioma-like and lymphangiectatic KS.[8]

The KS usually starts as a bluish-red macule on the distal portions of the lower extremities and the lesions progress slowly and may coalesce to form large plaques and may progress to form firm, nodular and fungiform tumors or may erode and ulcerate becoming amenable for FNAC testing. Histologically, there is dermal proliferation of interlacing bundles of spindle cells and intimately related, poorly defined slit-like vessels containing red blood cells. Mitotic activity is moderate and pleomorphism is usually absent. This lesion shows admixture of lymphocytes, hemosiderin-laden macrophages and plasma cells.

We conclude that though traditional biopsy testing is generally needed for confirmation, however FNAC has several advantages over the former including patient compliance, ease of procedure, no recurrences, and safety in immuno-compromised patients. Ancillary studies can be done on aspirates by cell-block technique. Also, PCR amplification techniques can be performed on cytological smears confirming detection of HHV-8 infection.

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