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Case Report

EBV-Associated Smooth Muscle Tumor Mimicking Schwannoma In AIDS Patient: A Case Report

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Abstract

Many tumors are associated with acquired immune deficiency syndrome (AIDS), such as Kaposi sarcoma, Burkitt's lymphoma, nasopharyngeal carcinoma, and smooth muscle tumor. This paper describes the Epstein-Barr virus (EBV)-associated smooth muscle tumor (EBV-SMT) in patient with AIDS, clinically mimicking schwannoma. So, EBV-SMT should be included in the differential diagnosis for a mesenchymal tumor arising in any organs in AIDS patients.

Introduction

Epstein–Barr virus (EBV) is one subtype of the human herpes virus. It is the cause of infectious mononucleosis and also associated with other neoplasms, such as Kaposi sarcoma, Burkitt's lymphoma, nasopharyngeal carcinoma, as well as smooth muscle tumor [1,2]. This virus spread via the droplet of saliva during acute infection and can establishes latent infection in B-cell. Recently, some authors claimed that there is strong supportive evidence between EBV and some tumors, but these tumors arise in cell types which no latency biological equivalent has been found. Examples of these tumors are nasopharyngeal carcinoma, gastric carcinoma, salivary gland carcinoma, and also smooth muscle tumor [3].

Human Immunodeficiency Virus (HIV) is a retrovirus which can spread by blood product, sexual activities, and also feto-maternal transmission. This virus entry into T-lymphocytes and impair the cell-mediated immunity, causing AIDS which is pandemic problem nowadays. Regarding to central nervous system (CNS), many disorder can produce by HIV infection, such as direct effect by HIV, opportunistic infection by other agents, and systemic non-specific manifestations by metabolic derangement [3]. Therefore a differential diagnosis of CNS mass lesions in AIDS patients is clinically difficult.

Case Report

A 26 year-old male, referred to Prasat Neurological Institute, during July 2017 – February 2018, with clinical history of seropositive status, treated with Zidovudine, Lamivudine, Efavirenz for 2 years. The recent CD4 cell count was 138 cells/mm^3. Deny of underlying disease, alcoholic drinking, smoking and drug allergy, but presence of history of illicit drug using (Amphetamine, Marijuana). He has presented with paraplegia of lower extremities for 3 months. The motor powers were grade 0 at lower extremities, but grade IV for upper extremities. Urinary sphincter tone was loose. Cranial nerve, eye and other physical examinations were within normal limit. Blood chemistry and chest x-ray were normal. He developed paraplegia of lower extremities for 3 months. The magnetic resonance imaging (MRI) showed intradural-extramedullary mass at C6 with severe cord compression (Figure 1).

Pathology revealed compact spindle cells with vesicular nuclei and eosinophilic cytoplasm. Mitotic figures were 10/10 High Power Fields, but necrosis was not found. The tumor cells were positive to smooth muscle actin (SMA) but negative to epithelial membrane antigen (EMA), S-100, Desmin, STAT-6, and TLE1. In situ hybridization for EBV-encoded ribonucleic acid (EBER) revealed positive reaction in almost the entire tumor cells (Figure 2).

Ziehl-Neelsen stains showed none of aid-fast bacilli. The patients had mild fever and pain postoperative. Few days later he was sent back home without any complications.

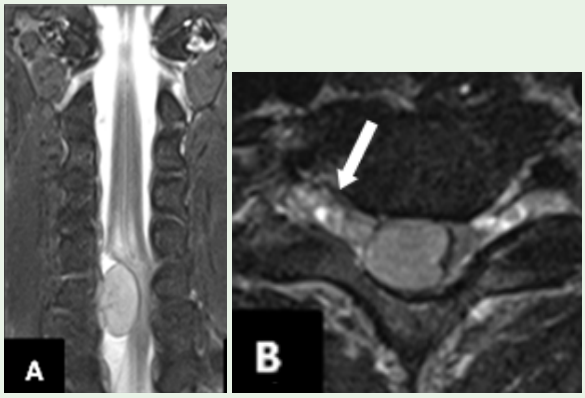

Figure 1: MRI of C-spine. (A) Axial T2-weighted MRI with fat suppression showing an intradural-extramedullary mass at right side of C6 level with compressive myelopathy at the adjacent cervical cord. (B) Coronal T2-weighted MRI with extension of the mass into right C6-7 neural foramen (white arrow).

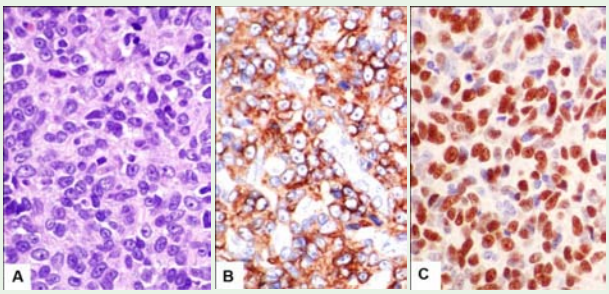

Figure 2: (A) Histopathology of the specimen showing compact spindle cells with cigar-shaped nuclei. Noted that prominent nucleoli are also seen (H&E, 100X). (B) Immunohistochemical stain for SMA (400X). (C) In situ hybridization for EBER (400X).

Discussion

Since there is increasing rate of AIDS cases, the numbers of EBV-SMT continue to grow. Some authors found that multiple lesions are unusual in immunocompetent hosts. And the most common location is CNS, following by soft tissue, lung, liver, colon, adrenals, hematopoietic system (spleen and lymph nodes), and bone. Regarding to the CNS, EBV-SMT can be dural, epidural or extradural [1,4,5]. Suankratay et al reported the EBV-SMT cases from Thailand since 2001-2003, 4 out of 9 cases were intraspinal masses, and all of them were epidural in origin. Moreover, 2 of 4 epidural lesions, the masses extended into the spinal canal through the neural foramen and showed enhancing dumbbell-shaped features by magnetic resonance imaging (MRI) [6]. In our patient, the tumor was found in the spinal cord, and located in intradural- extramedullary space. We believed that this is creeping-extending part of the tumor through neural foramen. These finding is compatible with nerve-sheath tumor and should be listed in the differential diagnosis.

Histologically, schwannoma consist of the spindle cells with wavy palisading nuclei and also are strongly reactive to S-100. While tumor cells of SMT show cigar-shaped nuclei and sometimes contain prominent nucleoli. They are usually reactive to SMA, but negative to S-100. neurofibroma shows small wavy nuclei with variable myxoid/collagenous background. The immunohistochemical tests of neurofibroma are similar to schwannoma (reactive to S-100). Meningioma is medium-sized spindle cells which characterized by indistinct cytoplasmic border and oval nuclei with occasionally intranuclear inclusion. Whorling of neoplastic cells is usually seen. This tumor is reactive to EMA, but non-reactive to markers of smooth muscle.

Another entity that could be seen in immunocompromised individuals is “mycobacterial spindle cell pseudotumor”. This lesion reveals numerous acid-fast bacilli within the spindle cells [7-9]. In our cases, the spindle cells do not demonstrate any acid fast bacilli by Ziehl-Neelsen stain.

Regarding to the prognosis, EBV-SMT has more favorable outcome, compared to conventional leimyosarcoma that often progresses with hematogenous spread and distant metastasis [1,10]. However, some researchers found that the histologic features of EBV-SMT did not correlate well with the clinical outcome [6]. In summary, EBV-SMT should always be included in the differential diagnosis for a mesenchymal tumor arising in any organs in AIDS patients of all ages.

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