Journal of Clinical Neuroscience xxx (2018) xxx–xxx

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Case report

Epstein-Barr virus-associated smooth muscle tumor involving the spine of an HIV-infected patient: Case report and review of the literature

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ARTICLE INFO

Article history:
Received 20 December 2017
Accepted 12 March 2018
Available online xxxx

Keywords:
Epstein-Barr virus
Smooth muscle tumor
Spine
HIV
Rare
Case report

ABSTRACT

Within the last two decades, there have been multiple reports of Epstein-Barr virus (EBV)-associated smooth muscle tumors in immunocompromised patients. This includes HIV-infected patients, post-transplant patients, and patients with congenital defects of their immune systems. Here we report the case of a 24-year-old African American female with congenital HIV presenting with progressive lower extremity weakness, constipation, aching pain in her shoulders, and subcostal anesthesia. Magnetic resonance imaging (MRI) revealed a large circumferential lesion extending from T1-T3 and a smaller left paraspinal lesion at C6-C7. The T1-T3 mass was excised via a right-sided costotransversectomy with laminectomy and fusion from T1-T3. Highly active antiretroviral therapy (HAART) was started postoperatively, and adjuvant radiotherapy was initiated but patient was lost to follow-up. Surgical pathology demonstrated a smooth muscle tumor diffuse nuclear positivity for EBV-encoded small RNA 1 by *in situ* hybridization. Although eight studies have reported HIV patients with EBV-associated smooth muscle tumors of the spine, to the author's knowledge, this is the first review comprised solely of patients with spinal involvement with the addition of our patient case.

1. Introduction

Within the last two decades, there have been multiple reports of Epstein-Barr virus (EBV)-associated smooth muscle tumors (SMTs) in immunocompromised patients. This includes HIV-infected patients, post-transplant patients, and patients with congenital immunological defects. In HIV-positive cases, patients are most commonly young adults with congenitally-acquired HIV infections and present with tumors in any of a variety of locations. Most commonly, these lesions are observed in the genitourinary or gastrointestinal tracts; however previous reports have documented spinous pathology [1]. Due to the rarity of EBV-associated SMTs, there is no established standard of care or tumor grading system. Hererin, we present a rare case of an HIV-infected patient with a primary EBV-associated SMT of the spinal column treated with surgical resection.

2. Case report

A 24-year-old African American female with a history of congenital HIV infection (off antiretroviral therapy for > 5 years) presented to the emergency room after one month of progressive lower extremity weakness, constipation, aching pain in bilateral shoulders, and numbness below T8 dermatome.

An MRI demonstrated a T1-T3 circumferential lesion extending through the T2-T3 right neural foramina and compressing the spinal cord (Fig. 1). A small lesion was also observed in the left paraspinal soft tissues extending through the left C6-C7 neural foramen, however, it demonstrated no evidence of neural element compression. The patient underwent a T1-T3 laminectomy for decompression and tumor excision, which was followed by stabilization with T1-3 pedicle screw fixation for stabilization (Fig. 2). She was subsequently admitted to the AIDS inpatient unit, where she started on elvitegravir-cobicistat-emtricitabine-tenofovir alafenamide for her HIV infection (Absolute CD4 count, 2 cells/mm^3; HIV RNA level, 292,000 copies/mL; no evidence of drug resistance). The patient also initiated adjuvant radiotherapy for the residual tumor, but was lost to follow-up before the course could be completed.

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https://doi.org/10.1016/j.jocn.2018.03.009
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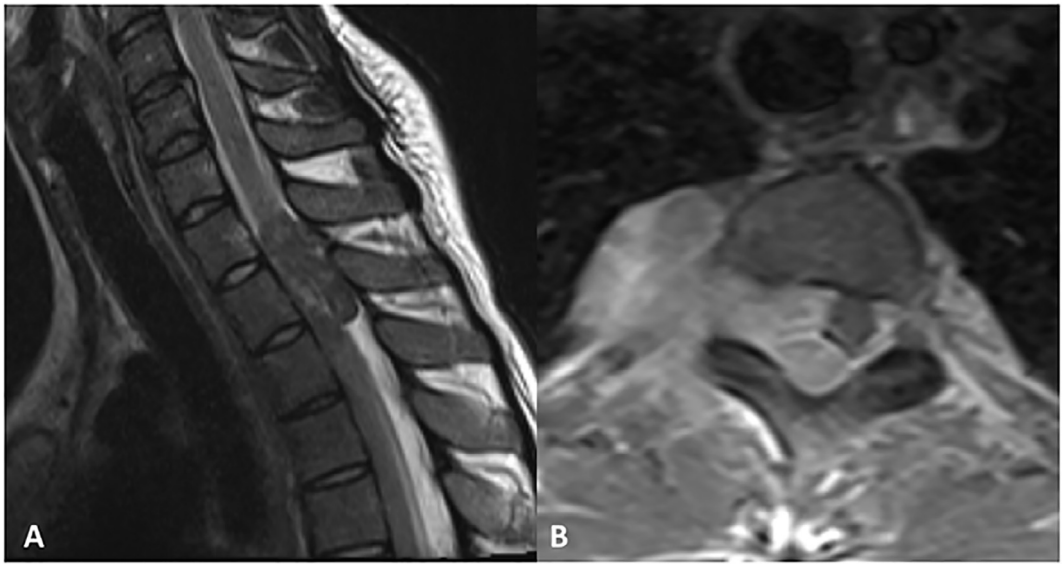

Fig. 1. Preoperative imaging demonstrating a right paraspinal soft tissue mass extending into the neural foramina from T1-T3, leading to extradural spinal cord compression and leftward shift. A. Sagittal T2-weighted MRI. B. Axial T1-post contrast MRI at the T2 vertebral body level.

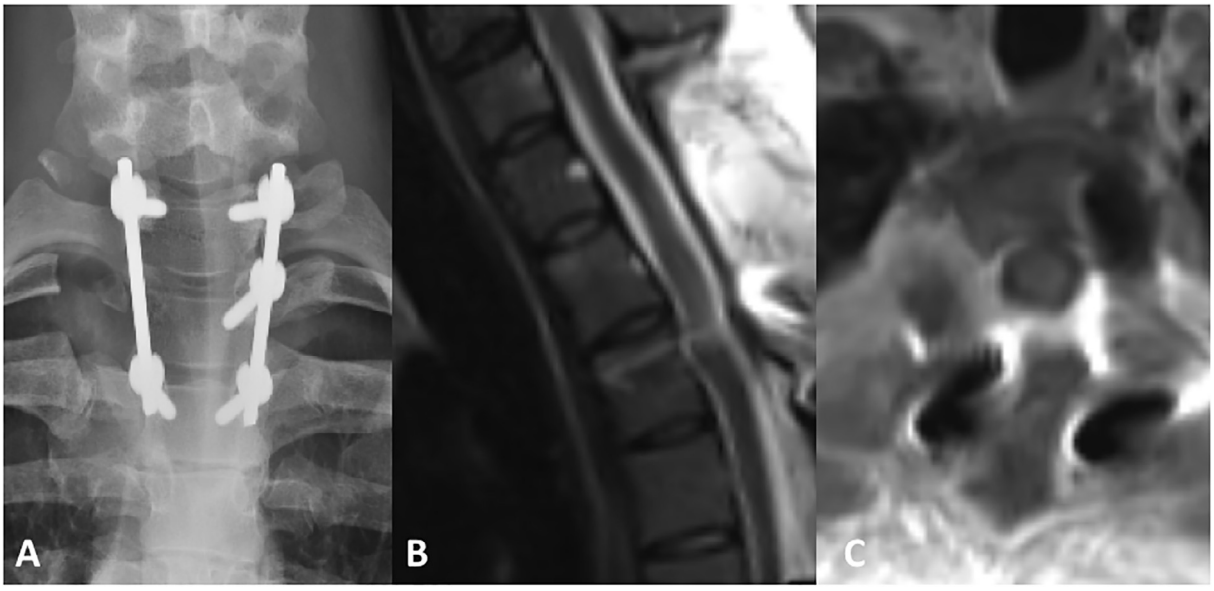

Fig. 2. Post-operative imaging demonstrating spinal cord decompression and posterior instrumentation from T1-T3 via a right-sided costotransversectomy approach. A. AP radiograph B. Sagittal T2-weighted MRI C. Axial T1 post-contrast MRI at the level of the T2 vertebral body.

A tumor specimen obtained during the surgery was sent for pathological analysis. The specimen was found to stain positive for desmin, smooth muscle actin (weak), collagen IV, p16, p53 (scattered), and S-100 protein (focal), and negative for SOX-10 and CD34. Although her quantitative polymerase chain reaction (PCR) for plasma EBV DNA was negative (limit of detection 50 EBV genome copes), *in situ* hybridization for EBV-encoded small RNA 1 (EBER1) was diffusely positive in the tumor. A Ki-67 immunostain demonstrated focally increased proliferation index. A mitotic count yielded low mitotic activity. Histopathological slides are seen in Figs. 3 and 4.

3. Discussion

Epstein-Barr virus (EBV)-associated smooth muscle tumors (SMTs) have been reported throughout the last four decades in immunodeficient patients [1,2]. The first reported case of an SMT in the presence of human immunodeficiency virus (HIV) was described in 1990 by Chadwick et al. who observed leiomyomas and leiomyosarcomas in the lungs and gastrointestinal (GI) tracts of three HIV-infected children [3]. Five years later, Lee et al. reported similar tumors composed of smooth muscle in the lungs, livers, and GI tract of three children who were being immunosuppressed after organ transplants [4]. After *in situ* hybridization, they found clonal EBV DNA in all tumor specimens, leading to the first association of EBV with SMTs in immunosuppressed patients [4]. These studies suggested a link between SMTs and EBV in patients with inhibited immune systems [4,5]. A subsequent study by Dekate and Chetty found that the EBV-associated SMTs occur most often in HIV-positive patients, similar to our current patient, followed by immunosuppressed post-transplant patients, and lastly, patients with congenital immunodeficiencies other than HIV [1].

Please cite this article in press as: Ehresman JS et al. Epstein-Barr virus-associated smooth muscle tumor involving the spine of an HIV-infected patient: Case report and review of the literature. J Clin Neurosci (2018), https://doi.org/10.1016/j.jocn.2018.03.009