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Primary hepatic Epstein–Barr virus-associated CD30-positive peripheral T-cell lymphoma of cytotoxic phenotype

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ABSTRACT

Primary hepatic peripheral T-cell lymphoma (PTCL) is exceedingly rare. We encountered such a case in a 58-year-old Hispanic female with a history of chronic sinusitis and hypothyroidism who presented with 4 weeks of fever and weight loss. Laboratory studies showed altered liver function and mild pancytopenia. Hepatitis and HIV infection were excluded by negative serological tests. A computed tomography (CT) scan showed innumerable small low-density lesions throughout the liver without splenomegaly or lymphadenopathy. CT-guided liver core biopsy showed scattered small lymphoid aggregates located mainly in the portal tracts and periportal regions. Within the lymphoid aggregates, scattered large pleomorphic lymphoma cells were seen, admixed with smaller lymphoid cells and histiocytes. By immunohistochemistry, the lymphoma cells expressed CD2, CD3, CD8, CD30, CD43, CD45, granzyme B, TIA-1, and negative for CD4, CD5, CD7, CD56, βF1, ALK-1, and B-cell markers. In situ hybridization for Epstein–Barr virus-encoded RNA (EBER) was positive in some lymphoma cells. To our knowledge, this is the first reported case of primary hepatic Epstein–Barr virus-associated PTCL with CD30 expression.

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1. Introduction

Primary hepatic lymphoma is a very rare malignancy. It constitutes about 0.4% of all primary extranodal non-Hodgkin lymphomas (NHLs) and 0.016% of all cases of NHLs (Freeman et al., 1972). The largest series of patients with primary hepatic lymphomas reported in the literature included 24 patients, in which 78% were identified as diffuse large B-cell lymphoma (Page et al., 2001). Other less common histological types of primary hepatic lymphomas described in the literature include follicular lymphoma, MALT lymphoma, mantle cell lymphoma, and other B-cell lymphomas (Page et al., 2001). Primary hepatic peripheral T-cell lymphoma (PTCL) is exceedingly rare, with less than 20 cases reported in the literature to date (Andreola et al., 1988; Anthony et al., 1990; Lei et al., 1995; Kim et al., 2000; Schweiger et al., 2000; Stancu et al., 2002; Leung et al., 2009; Miyashita et al., 2011; Mishra et al., 2013; Hu et al., 2014). Due to the scarcity of this entity, well-defined clinical and pathological features, prognostic factors, and standard management are difficult to determine. Therefore, continuous reporting of primary hepatic PTCL will bring to the awareness of the clinical and pathological spectrum of the disease, improve diagnostic accuracy, and provide better clinicopathologic correlation and perhaps better management of this tumor. In the present study, we report a unusual case of autopsy-proven primary hepatic PTCL in a middle-aged female.

2. Case report

A 58-year-old Hispanic female patient originally from Mexico, who has a history of chronic sinusitis and hypothyroidism, presented to Harbor-UCLA Medical Center with 4 weeks of persistent fever, chills, cough, nausea, poor appetite, and weight loss. The patient denied shortness of breath, chest pain, abdominal pain, vomiting, diarrhea, and dysuria. The general physical examination was unremarkable with no palpable cervical, axillary, or inguinal lymphoadenopathy. The laboratory studies on admission included a white blood cell count of 3.2 K/mm^3 (reference range 4.0–10.0 K/mm^3; 61.5% neutrophils, 36.3% lymphocytes, 2.1% monocytes, and 0.1% basophil), a hemoglobin concentration of 12.7 g/dL (reference range 11.9–14.9 g/dL), and a platelet count of 220 K/mm^3 (reference range 150–420 K/mm^3). Blood chemistry tests were normal except for a decrease in sodium level at 123 mmol/L (reference range 136–144 mmol/L), a decrease in chloride level at 93 mmol/L (reference range 101–111 mmol/L), a decreased in calcium level at 7.9 mg/dL (reference range 8.9–10.3 mg/dL), and a mild elevation in creatinine at 1.22 mg/dL (reference range 0.6–1.1 mg/dL). Liver function tests showed a total bilirubin of 1.1 mg/dL (reference range 0.3–1.2 mg/dL), a direct bilirubin of 0.5 mg/dL (reference range 0.1–0.5 mg/dL), an elevated aspartate aminotransferase (AST) level of 695 U/L (reference range 15–41 U/L), an elevated alanine aminotransferase (ALT) level of 477 U/L (reference range 7–35 U/L), and an elevated alkaline phosphatase level of 418 U/L (reference range 38–126 U/L). The serum lactate dehydrogenase level was 980 U/L (reference range 98–192 U/L).

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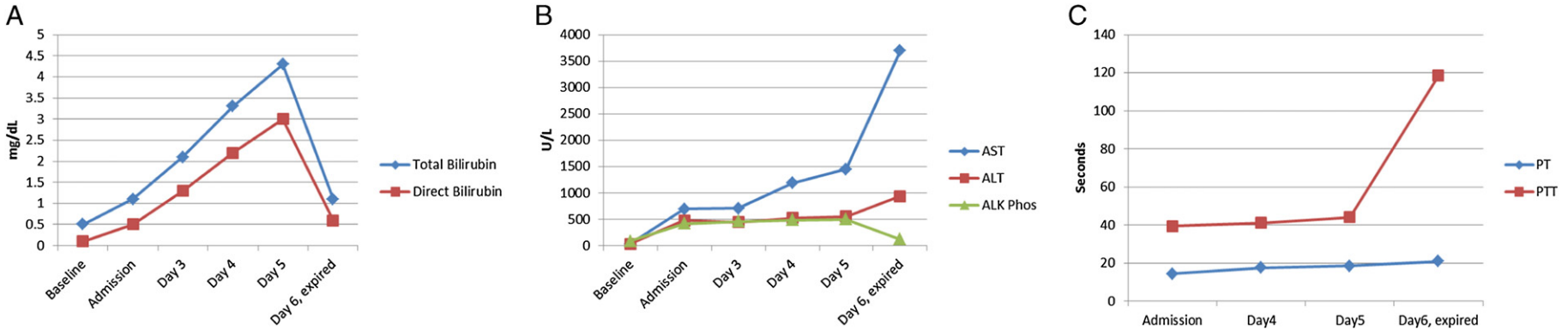

Fig. 1. The patient's liver function tests and coagulopathy profiles during the hospitalization course. (A) Total bilirubin and direct bilirubin levels, measured in mg/dL. (B) Alanine aminotransferase (ALT), aspartate aminotransferase (AST), and alkaline phosphatase levels, measured in U/L. (C) Prothrombin time (PT) and partial thromboplastin time (PTT) in seconds.