Clinical Imaging 39 (2015) 525–528

Contents lists available at ScienceDirect

Clinical Imaging

journal homepage: http://www.clinicalimaging.org

Multimodality imaging of Epstein–Barr virus-associated inflammatory pseudotumor-like follicular dendritic cell tumor of the spleen: case report and literature review

Pauline L. Bui [a,*,1], Rafael A. Vicens [b], Jason R. Westin [c], Corey T. Jensen [b,2]

[a] Department of Diagnostic Radiology, Baylor College of Medicine, One Baylor Plaza, Houston, TX 77030
[b] Department of Diagnostic Radiology, Unit 1473, The University of Texas MD Anderson Cancer Center, 1515 Holcombe Blvd., Houston, TX 77030–4009
[c] Department of Lymphoma/Myeloma, Unit 0429, The University of Texas MD Anderson Cancer Center, 1515 Holcombe Blvd., Houston, TX 77030–4009

ARTICLE INFO

Article history:
Received 17 August 2014
Received in revised form 5 December 2014
Accepted 24 December 2014

Keywords:
Inflammatory pseudotumor
Splenic pseudotumor
EBV
IPT
Epstein–Barr virus

ABSTRACT

Inflammatory pseudotumors (IPTs) are rare tumors of unknown etiology; however, there is a strong association with the Epstein–Barr virus (EBV). EBV-positive IPTs are typically found in the liver and spleen. While many EBV-positive splenic IPTs contain follicular dendritic cell (FDC) proliferations, they are not aggressive such as with conventional FDC tumors. EBV-positive splenic IPTs have been reported with low malignant potential. We present a case with multimodality imaging of an EBV-positive IPT-like tumor with FDC features.

1. Introduction

Inflammatory pseudotumor (IPT) is a rare benign tumor that can occur within various sites in the body. Approximately 80 cases of IPT in the spleen have been reported since the first described cases in 1984 [1]. The cause of IPT remains unknown; however, infectious or autoimmune causes are proposed mechanisms of pathogenesis [2]. One pathogen known to be associated with IPT is the Epstein–Barr virus (EBV). Interestingly, the EBV-positive subgroup of IPTs are predominantly found in the liver and spleen; this EBV-positive subgroup has also been shown to display low malignant potential with reports of recurrence after surgical resection [3], which is distinct from the typically benign conventional IPT. This different clinical behavior reflects the increasing understanding of histopathogenetic heterogeneity in IPTs and highlights the importance of improved radiologic and pathologic assessment. Authors have suggested that a spectrum of pathologic appearances between EBV-positive IPTs and IPT-like FDC tumors exists and that these tumors may represent the same entity [3]. Herein, we present one of only a few reported cases that include multimodality imaging of a proven EBV-positive IPT-like follicular dendritic cell (FDC) tumor of the spleen [4,3,2], which includes both magnetic resonance imaging (MRI) and fluorodeoxyglucose positron emission tomography ([^{18}F] FDG PET)-CT.

2. Case description

A 50-year-old woman presented with vague abdominal pain. Physical examination revealed mild tenderness to palpation in the left upper quadrant. Laboratory evaluation was significant only for elevated immunoglobulin levels at 1650mg/dl (normal range, 700–1600mg/dl). An abdominal computed tomography (CT) examination was obtained for further evaluation, which revealed a 6cm mass within the spleen containing a small calcification (Fig. 1a–c). The splenic lesion was further assessed with MRI and subsequently with [^{18}F] FDG PET-CT [Figs. 2a, 2b, 3a–c, 4]. The MRI depicted the mass to be iso- to hypointense relative to the spleen on precontrast T1- and T2-weighted sequences with varying degrees of heterogeneity [Fig. 2 and b]. Regions of serpiginous T1 hyperintensity were present throughout the mass on the precontrast, fat-saturated T1-weighted sequence. A central confluent area within the mass was T1 and T2 hypointense. No signal loss was identified between the in- and out-of-phase gradient echo sequences, and the contrast enhancement pattern was the same as that seen on CT [Figs. 2b, 3a–c]. The [^{18}F] FDG PET-CT [Fig. 4] revealed that the majority of the tumor to be hypermetabolic (maximum standardized uptake value: 8.5). The patient underwent a splenectomy, and the subsequent pathologic evaluation revealed an EBV-positive IPT with varying degrees of FDC features (Fig. 5 and b).

* Corresponding author. Department of Diagnostic Radiology, Baylor College of Medicine, One Baylor Plaza, Houston, TX 77030. Tel.: +214-820-9227.
E-mail addresses: pbui588@gmail.com (P.L. Bui), RVicens@mdanderson.org (R.A. Vicens), cjensen@mdanderson.org (C.T. Jensen).
[1] Present address: 3900 Junius St. Suite 100 Dallas, TX 75246.
[2] Tel.: +1 713 745 3835; fax: +1 713 745 1152.

http://dx.doi.org/10.1016/j.clinimag.2014.12.021
0899-7071/

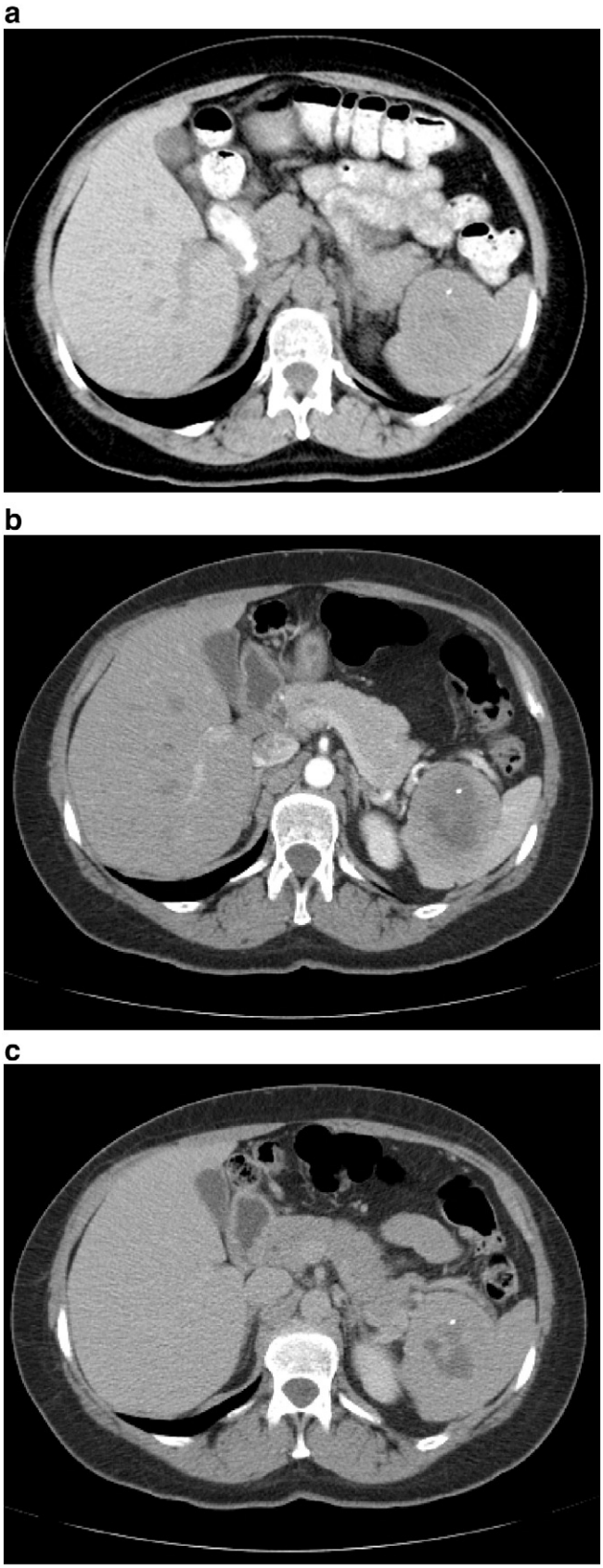

Fig. 1. (a–c): Axial CT of the abdomen shows a round, exophytic, splenic mass with a small internal calcification. The mass is nearly isoattenuating on precontrast imaging (a) with progressive enhancement from the late arterial (b) to the delayed phase (c).

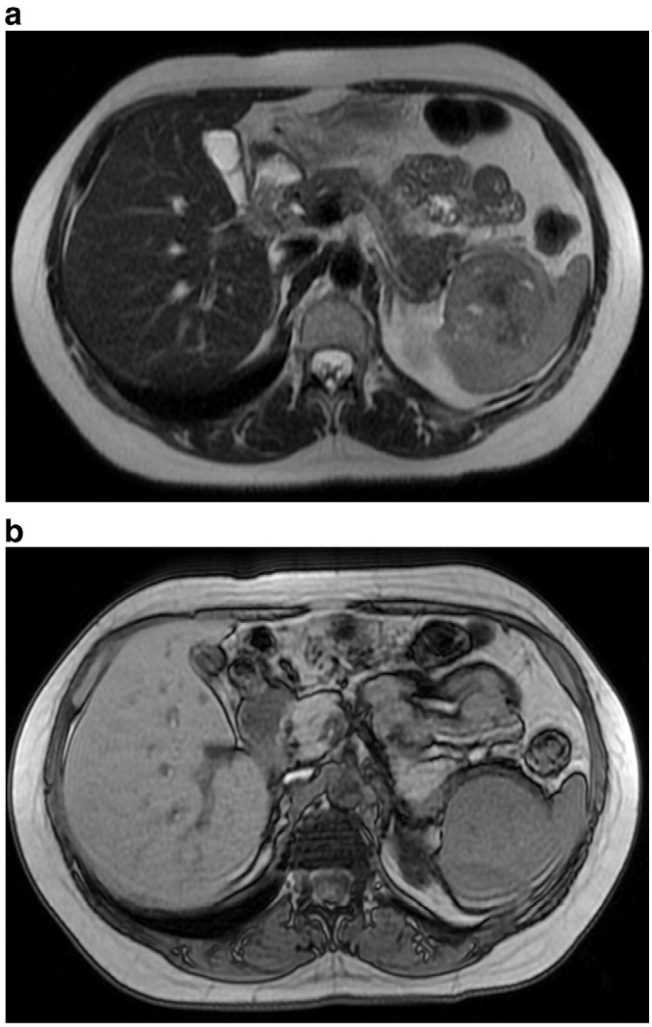

Fig. 2. (a) Axial T2-weighted single-shot fast spin echo abdominal MRI shows a predominantly isointense splenic mass with a hypointense center and multifocal areas of hyperintensity. (b) Axial out-of-phase Gradient Echo image shows no areas of signal reduction to indicate intracellular lipid within the splenic mass.

3. Discussion

EBV-positive IPTs are distinct from conventional IPTs in that they occur only in the spleen, liver, or lymph nodes. Previous reports have shown the presence of EBV in 40–67% of IPTs in the spleen and liver [5,6]. In contrast to their clinically benign EBV-negative counterparts, some EBV-positive IPTs are clonal neoplasms with low malignant potential [5]. Further details and questions surround the classification of the EBV-positive subgroup related to their observed degree of FDC differentiation. Kiryu et al. suggested that a spectrum of pathologic appearances between EBV-positive IPTs and IPT-like FDC tumors exists and that these tumors may represent the same entity [3].

Limited demographic assessment of EBV-positive IPTs has not indicated a distinct age range [6]. A study that included both EBV-positive IPTs and IPT-like FDCs showed a female predominance with a male-to-female ratio of 5:9 [1]. This is in contrast to the lack of gender predilection seen in FDC tumors that are not classified with IPT features [7].

Patients with EBV-positive IPTs or IPT-like FDCs in the spleen are often asymptomatic but may present with vague complaints such as epigastric pain or fever [6]. Many times the splenic mass is an incidental finding on imaging obtained for other indications [5]. Symptoms associated with lymphoma, such as weight loss and night sweats, have also been reported with IPTs [8,6].