Available online at www.sciencedirect.com
ScienceDirect
Annals of Diagnostic Pathology 11 (2007) 127–131
Annals of DIAGNOSTIC PATHOLOGY

A case of primary unilateral adrenal Burkitt-like large cell lymphoma presenting as adrenal insufficiency

Ling Zhang, MD[a], Sameer S. Talwalkar, MD[a], Saad Paul Shaheen II, MD[b,*]

[a]*Department of Pathology and Laboratory Medicine, University of Louisville, Louisville, KY 40202, USA*
[b]*Veterans Affairs Medical Center, University of Louisville, Louisville, KY 40206, USA*

Abstract Primary adrenal lymphoma is extraordinarily rare, in comparison with secondary adrenal involvement by non-Hodgkin lymphoma. Although higher-resolution imaging techniques have enhanced detection of adrenal masses, biopsy or excision is often needed for definitive diagnosis. Percutaneous computed tomography–guided fine needle aspiration has great diagnostic value in the workup of adrenal masses, but is limited by sampling error and artifacts. Primary adrenal lymphoma most commonly manifests with diffuse large B-cell morphology. Burkitt-like large cell lymphoma morphology has been previously reported only once, to our knowledge. We report an 80-year-old man presenting with unilateral primary adrenal lymphoma showing Burkitt-like morphology and adrenal insufficiency. Fine needle aspiration yielded a dispersed population of monomorphic, medium to large cells suggestive of lymphoma. Although dispersed cell populations cytologically favor lymphoma, metastatic poorly differentiated carcinoma and adrenal cortical carcinoma can manifest similarly. Integrated histological, immunohistochemical, and flow cytometric immunophenotyping would provide an accurate and definitive diagnosis. We review the literature and discuss important issues with regard to diagnosis.

Keywords: Large cell lymphoma; Adrenal insufficiency; Fine needle aspiration; Burkitt's lymphoma

1. Introduction

Primary adrenal lymphoma (PAL) is defined as lymphoma arising within and confined to adrenal glands, in the absence of lymphadenopathy and leukemic blood picture [1-5]. Most patients with PAL present with bilateral disease, most of whom with adrenal insufficiency secondary to destruction of more than 90% of adrenal parenchyma by disease [1,2,5-7]. We describe a patient with unilateral PAL, who developed adrenal insufficiency. To our knowledge, unilateral PAL has not been associated with adrenal insufficiency [1,8]. Most PALs are of B-cell origin, most of which are diffuse large B-cell lymphomas (DLBCLs) [1,2,5-9]. Herein, we report a histological pattern that has been previously reported only once before our case [7].

* Corresponding author. Department of Pathology and Laboratory Medicine (113), Veterans Affairs Medical Center, Louisville, KY 40206, USA. Tel.: +1 502 895 3401x5534; fax: +1 502 894 6265.
E-mail address: spshaheen@pol.net (S.P. Shaheen).

1092-9134/$ – see front matter © 2007 Elsevier Inc. All rights reserved.
doi:10.1016/j.anndiagpath.2005.12.009

2. Case report

An 80-year-old white man with previous radical prostatectomy for prostatic adenocarcinoma and gastrectomy for peptic ulcer disease was seen in April 2003 for lower abdominal pain, anorexia, and cachexia. His medical problems included hypertension, peripheral vascular disease, and chronic obstructive pulmonary disease. He was found to have partial small intestinal obstruction on plain abdominal x-ray. Abdominal computed tomographic (CT) scan revealed an incidental homogenous soft tissue mass measuring 8 × 6.5 × 3 cm and entirely replacing left adrenal gland (Fig. 1). Five months later, his symptoms progressed to increased weakness, continued weight loss, and ambulatory difficulty. Physical examination revealed left abdominal tenderness, without palpable hepatosplenomegaly or lymphadenopathy. Initial laboratory studies revealed sodium level of 130 mmol/L (reference range, 133-150) and normal potassium and calcium levels. Intravenous gadolinium-enhanced magnetic resonance imaging revealed significant enlargement of left adrenal mass (12 ×

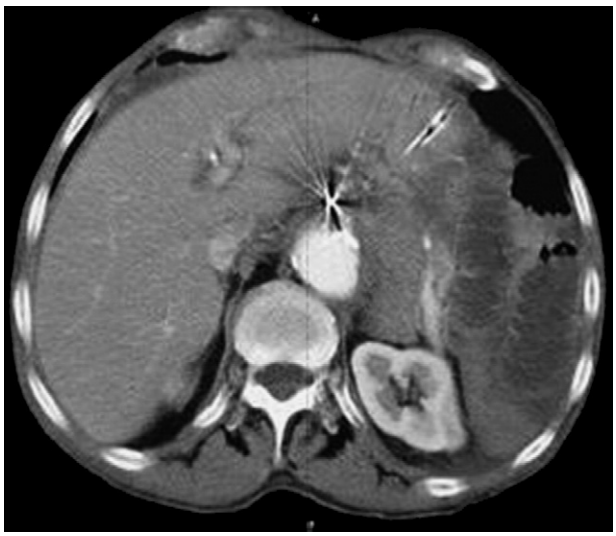

Fig. 1. Computed tomography of abdomen, demonstrating a nonenhancing, homogenous mass (5.5 × 4.5 cm) filling the adrenal and situated anteromedial to right kidney.

11 × 9 cm), which demonstrated a homogenous signal intensity with minimal enhancement and no evidence of necrosis. Vanillylmandelic acid, metanephrine, and normetanephrine levels were normal, excluding primary adrenal medullary neoplasm. Dexamethasone suppression test was negative, excluding primary adrenal cortical neoplasm. Corticotropin (ACTH) level was 5 pg/mL (reference range, 6-48 pg/mL) and cortisol level was 5.2 μg/dL (reference range, 3.1-16.66 μg/dL). Thyrotropin level was 2.25 μIU/mL (reference range, 0.35-5.5 μIU/mL). Dihydroepiandrosterone level was less than 15 μg/dL (reference range, 28-175 μg/dL). Rapid cosyntropin stimulation test, using 250 μg of intravenous cosyntropin (analog of ACTH), demonstrated plasma cortisol measurements of 10.8 and 6.74 μg/dL at 30 and 60 minutes, respectively. This incremental cortisol increase after cosyntropin injection was less than an adequate response of adrenal gland to cosyntropin stimulation (reference range, at least 7μg/dL at 0 minutes, 16 μg/dL at 30 minutes, and 18 μg/dL at 60 minutes poststimulation), strongly suggesting adrenal insufficiency.

3. Pathological findings

Under CT guidance, a fine needle aspiration of left adrenal lesion was performed. Material procured from this fine needle aspiration was smeared onto slides and stained by hematoxylin and eosin (H&E) and Papanicolaou methods. Unfortunately, air-dried smears stained with Diff-Quik were nondiagnostic (mostly blood). Smears showed a low number of dispersed medium to large lymphoid cells and stripped nuclei in a bloody background. No adrenal parenchymal cells or Reed-Sternberg cells were seen. The cell block, stained by H&E, showed a similar dispersed population of abnormal lymphoid cells at low and high powers, with both "stripped" lymphoid nuclei and

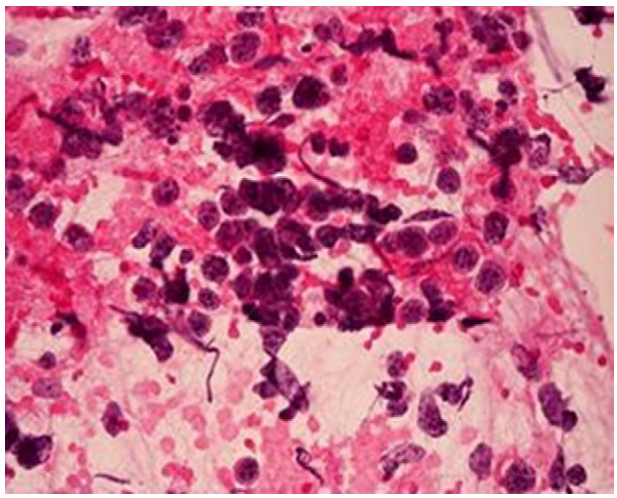

Fig. 2. Cell block of aspirated material from left adrenal (H&E, original magnification ×200).

atypical lymphoid cells with narrow cytoplasmic rims. Nuclear shape varied from ovoid to irregular and mildly pleomorphic (Fig. 2). Nuclear streaming and crush artifact were noted. Fine nuclear detail was not apparent. No vacuoles representing lipid-rich cytoplasm were apparent. Although lymphoma was suspected, resection for definitive diagnosis was performed. The patient underwent left adrenalectomy, splenectomy, distal pancreatectomy, and partial diaphragm resection for left adrenal tumor that entirely replaced adrenal gland and invaded into surrounding tissues. The left adrenal tumor weighed 916 g and measured 15.0 × 11.0 × 6 cm. Grossly, tumor demonstrated a fish flesh–like, tan-gray cut surface with focal punctate hemorrhages, partial encapsulation, and fine peripheral lobulation (Fig. 3). Histological sections showed a relatively monotonous and diffuse population of medium to large lymphoid cells with "squared-off" cytoplasmic borders, increased mitosis and apoptosis, and interspersed tingible

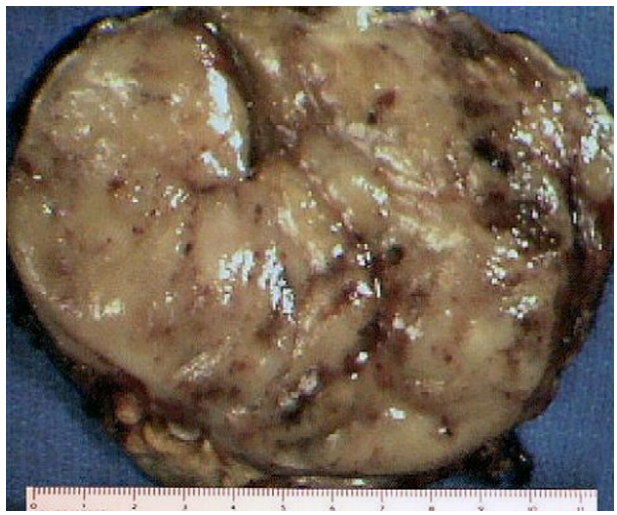

Fig. 3. Gross photograph of left adrenal, entirely replaced by a homogenous mass, with a fish flesh–like cut surface and ill-defined lobulation.