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Unilateral Primary Adrenal B-Cell Lymphoma Clinically Mimicking Chronic Gastritis

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Background:	Primary adrenal lymphomas are a very rare type of extranodal lymphoma, and they usually are found bilaterally. Symptoms of the disease are variable and depend on the type of lymphoma and/or presence of adrenal insufficiency. Magnetic resonance imaging is the best radiologic modality for differentiating lymphomas from other adrenal malignancies; however, histopathology is considered as the gold standard method for diagnosing extranodal lymphomas.
Case Report:	We present a case of unilateral adrenal lymphoma that was initially diagnosed as an infectious disease and chronic gastritis, based on clinical and laboratory findings. Ultrasonography detected an adrenal mass, and magnetic resonance imaging excluded common lesions of the adrenal gland and showed lymphadenopathy around the major vessels of the abdomen. A percutaneous biopsy of the mass and bone marrow biopsy confirmed the diagnosis of primary adrenal lymphoma without bone marrow metastasis.
Conclusions:	Extranodal lymphomas can occur in almost every organ, and if nonspecific clinical findings are combined with the presence of a solid organ mass, lymphoma should be included in the differential diagnosis.
MeSH Keywords:	Adrenal Gland Neoplasms • Adrenal Insufficiency • Fever of Unknown Origin • Magnetic Resonance Imaging
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Background

Non-Hodgkin lymphomas can arise from tissues other than lymph nodes, in which case they are called extranodal lymphomas. The most common locations of extranodal lymphomas are gastrointestinal tract, central nervous system, and skin [1]. Adrenal gland masses are extremely common, and the majority of them are found incidentally. Most common benign solid lesions of the adrenal gland include adenoma, myelolipoma, and the majority of pheochromocytomas, whereas most frequent malignant lesions are metastases and adrenocortical carcinoma. The adrenal gland is an unusual site of primary extranodal non-Hodgkin's lymphoma, accounting for 3% of primary extranodal lymphomas [2]. Patients with bilateral primary adrenal B-cell lymphoma (PAL) develop adrenal insufficiency, but unilateral PAL

can be asymptomatic, thereby leading to delayed diagnosis [3]. Imaging studies are crucial in the management of PALs, since proper imaging can prevent from unnecessary surgery by displaying features of lymphoma [4]. In this report, we aimed to describe magnetic resonance imaging (MRI) features of unilateral PAL, before and after chemotherapy, in view of the current literature.

Case Report

A 42-year-old male presented to our outpatient clinic with a 10-month history of fever (38°C), night sweats, malaise, abdominal pain, and a minimal weight loss after a trip to Africa. Arterial pressure, heart rate, and respiratory rate were within normal range, and physical examination revealed no abnormalities. The patient had no

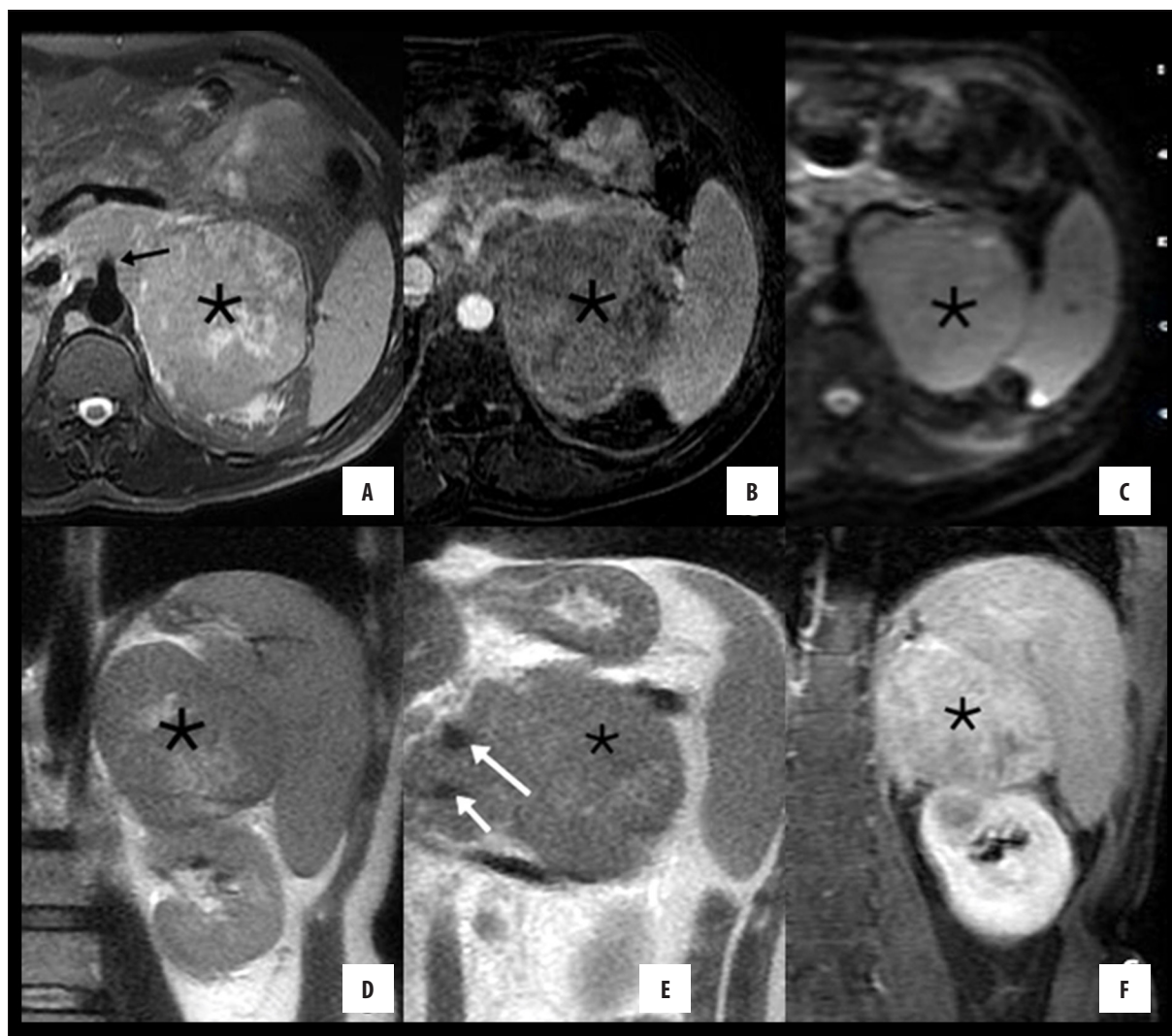


Figure 1. MRI was performed before chemotherapy treatment. Axial T2-weighted image (A) reveals a large left-sided adrenal mass (*) that extends anteriorly to the superior mesenteric artery (arrow). Axial (B) postcontrast subtraction MR images display a homogeneous contrast enhancement of the mass (*). On diffusion-weighted images, (C) the lesion showed restricted diffusion. On coronal T2-weighted images (D, E) and postcontrast subtraction MR (F) images, the mass enhances and displaces the left kidney inferiorly, the spleen laterally, and extends to the midline, encasing the celiac trunk and splenic vein (arrows).

history of malignancy, alcohol consumption, or smoking. There was leukocytosis and a high serum C-reactive protein (CRP) level, but serum electrolytes and creatinine were within normal limits. Serum cortisol and aldosterone levels were within normal limits, but serum lactate dehydrogenase (LDH) was elevated (455 UI/L, reference range 135–225 UI/L). An infectious disease was suspected, and the patient was treated with various antibiotics with no clinical remission. Subsequently, the patient underwent gastroscopy which indicated gastritis due to *Helicobacter pylori* infection, which was eradicated by proper treatment. However, symptoms did not respond well to treatment. Additionally, persisting left upper quadrant abdominal pain prompted us to perform abdominal ultrasonography which showed a 98×97 mm mass lesion on the left side of the abdominal cavity that pushed the left kidney inferiorly. To define the lesion more accurately, we performed abdominal contrast-enhanced MRI which revealed a solid mass arising from the left adrenal gland with homogeneous

contrast enhancement. Based on MRI features, adrenal adenoma and adrenal hemorrhage were excluded (Figure 1). The mass extended to the midline, encasing the celiac trunk and splenic vein. The pancreas and the spleen were displaced anteriorly, and the border with the left kidney could not be discriminated. The right adrenal gland was morphologically normal. There was paracaval and para-aortic lymphadenopathy. Lung cancer with adrenal gland metastasis was suspected, and thoracic computed tomography (CT) was performed. However, no parenchymal mass nor mediastinal lymph node enlargement were observed. An ultrasound-guided percutaneous tru-cut biopsy was performed to confirm the diagnosis. The patient was diagnosed with diffuse B-cell lymphoma by histopathology. Biopsy of bone marrow was normal, and metastatic disease was excluded. Finally, based on histopathological findings, the patient was diagnosed with PAL and referred for chemotherapy.



Figure 2. MRI after two courses of chemotherapy: Axial T2- weighted image (A), axial T1-weighted image (B), coronal T2-weighted image (C), and coronal postcontrast T1-weighted image (D) demonstrate a decrease in the size of mass (*).

After two courses of chemotherapy, MRI displayed regression of the mass (>50–60% of initial size) and a decrease in the number of enlarged lymph nodes (Figure 2). These results were interpreted as complete regression of the disease, and follow-up imaging was scheduled.

Discussion

PAL is an uncommon disease of the adrenal gland, with a male predominance (M/F ratio of 7/1). It constitutes less than 1% of all non-Hodgkin lymphomas and 3% of extranodal lymphomas [4]. It is usually found postmortem, and usually presents with bilateral adrenal masses [5]. Diffuse large B-cell lymphoma is the most common type of PAL, followed by mixed large and small cell, small noncleaved cell, and the undifferentiated types [6].

Histopathological confirmation of PAL is necessary, since management is different from other adrenal gland malignancies. Therefore, radiology is essential to guide the

management. The differential diagnosis of adrenal masses includes adrenal adenoma, hemorrhage, adrenal carcinoma, pheochromocytoma, adrenal cyst, tuberculosis, myelolipoma, and metastases of various cancers, such lung, breast, kidney, and pancreatic cancer or melanoma [7]. Adenoma has near-water attenuation on non-enhanced CT, and due to the high sensitivity of chemical shift MRI to the presence of small amounts of intravoxel fat, the majority of adenomas demonstrate signal intensity loss on opposed-phase GRE images. Thus, a drop in signal intensity greater than 20% is considered diagnostic for a lipid-rich adenoma [8]. For pheochromocytomas, the classic imaging feature is a “light-bulb” bright lesion on T2-weighted images and avid gadolinium enhancement on postcontrast images. The differential diagnosis of adrenal masses also includes tuberculosis, which usually shows symmetrical adrenal enlargement with central areas of low attenuation (caseous necroses), peripheral enhancement, and regional adenopathy on CT. In adrenal myelolipomas, fat attenuated components in the mass may be seen on CT. On MRI,

myelolipomas have high signal on T1-weighted images and signal loss on fat-suppressed sequences. Adrenal hemorrhage is usually a non-enhancing, homogenous, hyperdense lesion on non-contrast-enhanced CT [9]. On the other hand, adrenal lymphoma has variable imaging features. However, some of them should be kept in mind in particular; adrenal lymphomas usually present as a well-defined mass that displaces the adjacent structures but do not invade them. On CT, lymphoma usually looks like a hypodense, lobular, homogeneous, and slowly contrast enhancing solid mass; sometimes, it is accompanied by enlarged adjacent lymph nodes. On MRI, lymphomas tend to have low to intermediate signal intensity on T1-weighted images and moderately high signal intensity on T2-weighted images [10].

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Conclusions

PAL has remarkable imaging features that can help in diagnosis and management planning. When diagnosing an adrenal mass, PAL should be kept in mind in case of chronic nonspecific symptoms and an elevated serum LDH level.

Conflict of interest

The authors declared no conflicts of interest.