Radiology Case Reports 19 (2024) 158–163

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

Case Report

Bilateral primary adrenal diffuse large B cell lymphoma: A rare presentation ☆,☆☆

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ARTICLE INFO

Article history:
Received 7 July 2023
Revised 13 September 2023
Accepted 20 September 2023

Keywords:
Bilateral primary adrenal
CT scan
Lymphoma
Large B cell

ABSTRACT

Primary lymphoma of the adrenal gland is an uncommon origin of adrenal tumors; it must be explicitly invoked anytime bilateral adrenal affections are revealed. We report a case of bilateral primary adrenal diffuse large B cell lymphoma and perform a review of the literature. Our patient was a 55-year-old man who presented night sweats and a feeling of cardiac palpitations all evolving in a context of deterioration of his general condition. An ultrasound study was requested reporting a bilateral mass corresponding to the adrenal region. A thoraco-abdominopelvic CT scan was requested and revealed bilateral homogenous, polylobed, adrenal masses, discreetly enhanced after injection of contrast product. These masses were associated with multiple retroperitoneal, para-aortic, and celiomesenteric adenopathies. Anatomopathologic examination of the percutaneous CT-guided biopsy specimen of the adrenal tumor revealed the presence of diffuse large B cell lymphoma grade IIIB according to the Ann Arbor system. Primary adrenal lymphoma PAL on its own is an extremely rare disease entity and less than 100 cases have been reported in the last 40 years. A large proportion of PAL case reports showed that this disease usually has no excretory endocrine function and the symptoms are due to the pressure effect of the mass, whereas adrenal insufficiency usually exists. Our patient presented symptoms of indrenal insufficiency which seems to be the reason for the early diagnosis. Primary bilat-

☆ Acknowledgments: This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors. **Ethical approval for this study was not necessary**.

☆☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2023.09.064

eral adrenal lymphoma is very rare entity that should be kept in mind whenever bilateral adrenal masses are assessed in the CT scan images.

Introduction

There is a remarkable etiological diagnostic issue with the adrenal mass. Therefore, primary lymphoma of the adrenal gland is an uncommon origin of adrenal tumors; it must be explicitly invoked anytime bilateral adrenal affections are revealed [1,2]. Adrenal insufficiency may be the primary symptom of presentation, especially with bilateral involvement as in bilateral primary adrenal lymphoma [3]. The prognosis of primary adrenal lymphoma PAL is typically poor, and the 1-year survival rate is 17.5% [4]. We report a case of bilateral primary adrenal diffuse large B cell lymphoma and perform a review of the literature. The work has been reported in line with the SCARE criteria.

Case report

Our patient was a 55-year-old man with no medical history, who presented night sweats and a feeling of cardiac palpitations. On questioning, the patient reported also a deterioration of his general condition, with an asthenia and a weight loss of 7 kg over 1 month.

On general examination, we found a conscious patient respiratory stable and hemodynamically instable: the pulse rate was 113 bpm and the blood pressure was 98/77 mmHg. The patient was pale. On physical examination, no hemorrhagic syndrome or tumor syndrome were found, no peripheral adenopathy, and no splenomegaly or hepatomegaly. The rest of the clinical examination was normal.

On laboratory examination, the patient presents a regenerative anemia at 8.7 g/dL normochromic normocytic and a thrombocytopenia at 72,000 elements/mm^3. No abnormality was found on the other blood cells.

An ultrasound study was requested reporting a bilateral mass corresponding to the adrenal region. A thoracoabdominopelvic CT scan was requested and revealed bilateral homogenous, polylobed, adrenal masses, discreetly enhanced after injection of contrast product, measuring 130 × 103 × 91 mm and 114 × 91 × 78 mm in the right and left adrenal glands, respectively. These masses were associated with multiple retroperitoneal, para-aortic, and celiomesenteric adenopathies (Figs. 1 and 2).

Positron emission tomography with 18F-fluorodeoxyglucose (FDG-PET) showed bilateral hypermetabolic adrenal masses with SUVmax of 47 on the right and 50.2 on the left, with irregular contours (Fig. 3). There was also a supra-clavicular adenopathy, associated with bilateral hypermetabolic mediastinal adenopathies and external iliac hypermetabolic adenopathy.

Anatomopathologic examination of the percutaneous CT-guided biopsy specimen of the adrenal tumor revealed the presence of diffuse large B cell lymphoma grade IIIB according to the Ann Arbor system. The neoplastic cells were positive for CD20, MUM1 and BCL2. There was no expression for CD10, CD3, CD5, and BCL6. Ki-67 proliferation index was at 90%. Bone marrow biopsy did not show any marrow involvement.

During this period, the patient's condition deteriorated significantly, chemotherapy was immediately initiated once the diagnosis was found and the patient started four cycles of R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone) chemotherapy. The patient's general condition improved significantly. The thoracoabdominopelvic CT scan performed 1 month after the end of R-CHOP chemotherapy showed regression of the bilateral adrenal mass with minimal residual lesions. FDG-PET CT scan showed the regression of hypermetabolic adrenal masses as well as supra-clavicular, mediastinal, and external iliac adenopathy, 4 months after his initial admission at the hospital (Fig. 4).

Discussion

Less than 100 cases of PAL have been documented in the past 40 years, making it an extremely rare disease entity. It often presents with bilateral tumor masses (70%) and can results in complete or partial adrenal insufficiency [5,6].

Males are more predisposed with a male-to-female ratio around 1.8/1 [7]. Our patient was a male subject. The average age is 70 years old (range = 39-87) according to the existing literature [8]. In 85%-91% of cases of PAL, the diagnosis was diffuse large B cell lymphoma [9].

A large proportion of PAL case reports showed that this disease usually has no excretory endocrine function and the symptoms are due to the pressure effect of the mass,[7] whereas adrenal insufficiency usually exists. Our patient presented symptoms of indrenal insufficiency which seems to be the reason for the early diagnosis. Nevertheless, a few articles have reported normal adrenal function in B-PAL [10].

Age, tumor size, adrenal insufficiency, lactate dehydrogenase level, and performance status of the patient can significantly influence prognosis [11]. Survival time is short and a high degree of suspicion is needed in order to obtain a quick diagnosis.

Diagnosis can be established with the help of biopsy and histological examination [12]. In our case, we elected a percutaneous needle biopsy according to patient preference.

Histologic diagnosis is very important as the differential diagnosis of bilateral adrenal masses can vary from benign adrenal tumor to tuberculosis, primary adrenal lymphoma, metastatic malignancy, and pheochromocytoma.

Surgery, radiotherapy, and chemotherapy are available treatment options for PAL. Ideal management of PAL remains

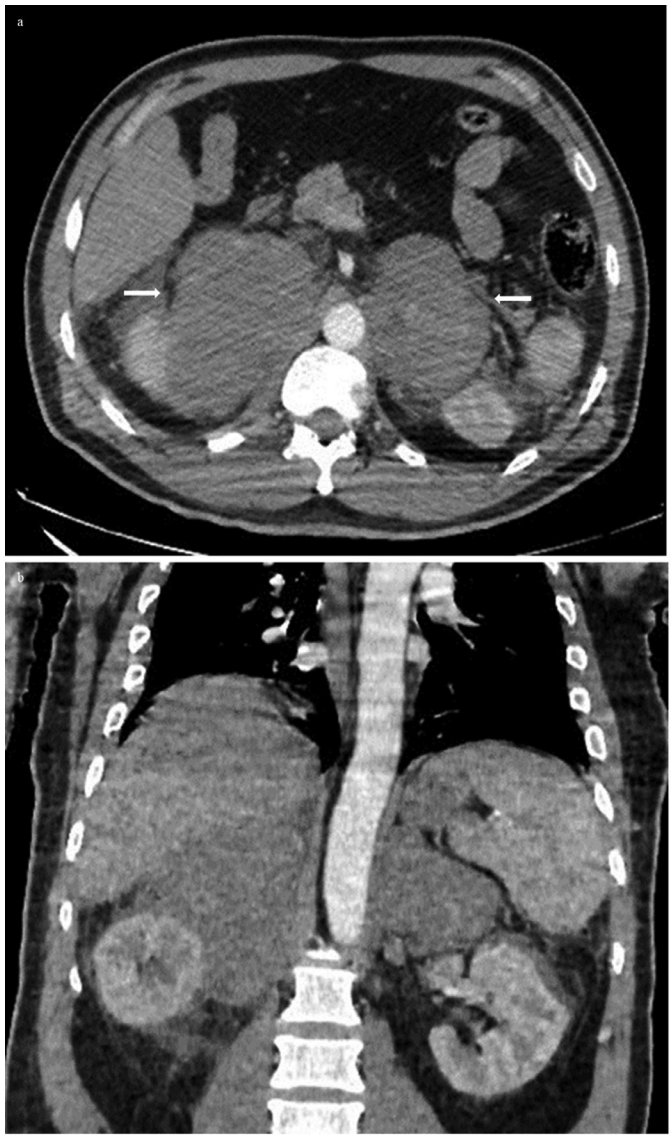

Fig. 1 – Postcontrast CT scan axial (A) and coronal (B) images showed bilateral adrenal masses which are homogenous with a slight contrast enhancement measuring 114 x 91 x 78 mm on the left (77.4 average HU), while the right adrenal gland measures 130 x 103 x 91 mm (73.7 average HU).

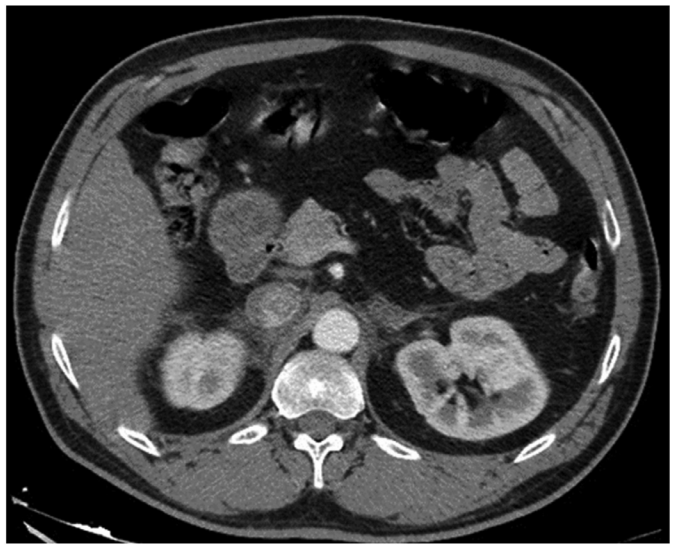

Fig. 2 – Postcontrast CT scan transverse view showing minimal bilateral residual lesions in the adrenal gland at the admission at hospital.

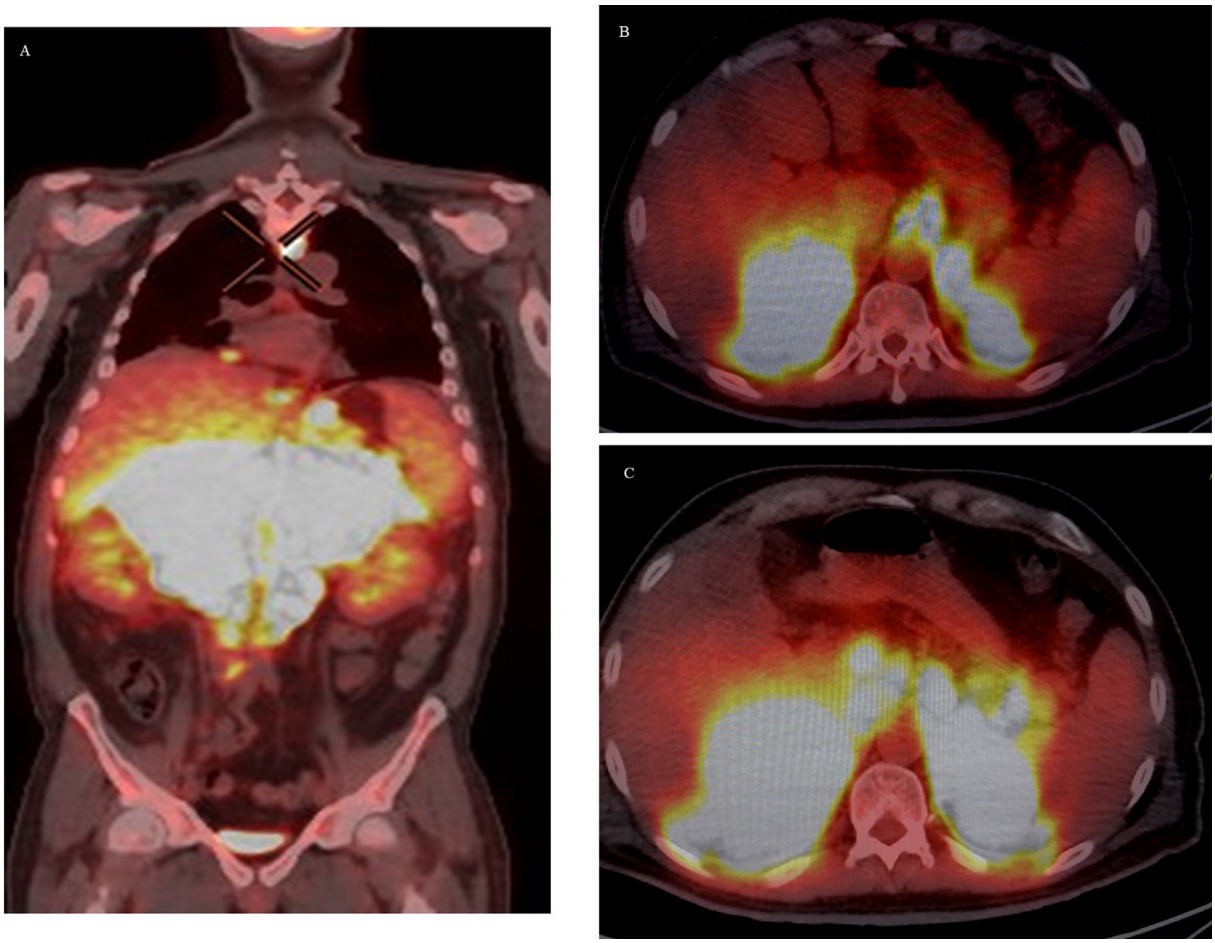

Fig. 3 – Positron emission tomography/computed tomography (PET/CT) coronal and axial views showing intensely hypermetabolic disease within the abdomen, involving both the adrenal glands 3 weeks after admission at the hospital.

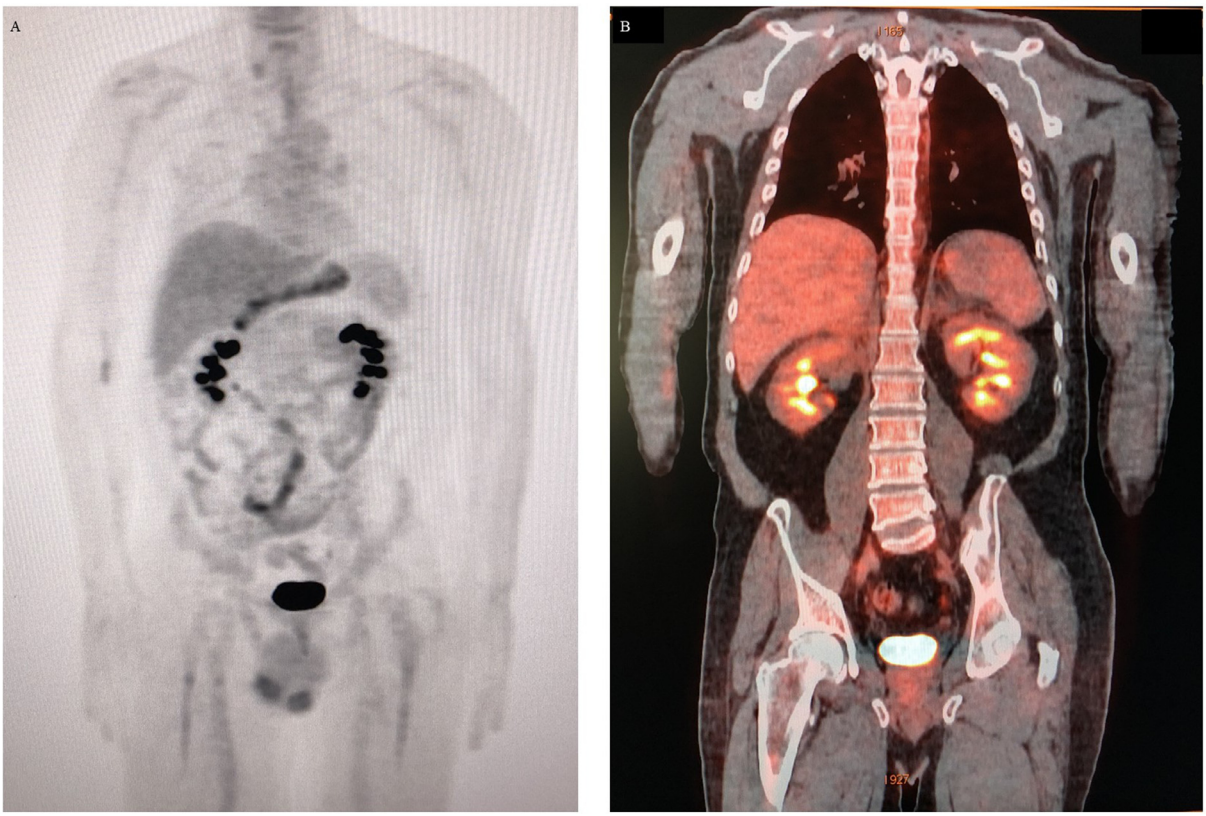

Fig. 4 – PET-CT coronal views showed the regression of hypermetabolic adrenal masses 4 months after his initial admission at the hospital.

controversial because PAL is an infrequent disease for which management options are supported by the few case series described in the literature. Adrenalectomy is not an adequate option. Currently, R-CHOP chemotherapy is the most effective treatment choice with respect to patient outcomes [13].

The combination of R-CHOP chemotherapy regimen has shown improvement in patients with diffuse large B cell lymphomas compared with CHOP alone [14].

Although the prognosis of PAL is very poor and complete remission with chemotherapy has been reported only in a few articles [15], 6 months after diagnosis and 1 month after R-CHOP chemotherapy termination, our patient was progressing well. It was, therefore, reassuring that post treatment scans showed regression of both adrenal masses.

We assume that this case will add to the few other PAL cases with adrenal insufficiency reported in the literature and demonstrated a reasonable response. However, more cases of PAL with adrenal insufficiency that responded well to chemotherapy are needed to be studied in order to identify the predictors of good clinical and radiological response to treatment.

Conclusion

Primary bilateral adrenal lymphoma is very rare entity. It affects mainly men but women are not excluded. This rare disease should be kept in mind whenever bilateral adrenal masses are assessed in the CT scan images. The diagnosis is essentially histological. The chemotherapy with or without radiotherapy is the usual treatment. The prognosis remains poor.

Patient consent

Written informed consent was obtained from the patient for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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