Case Report

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Symptomatic Adrenal Insufficiency due to Bilateral Adrenal Non-Hodgkin's Lymphoma

Bilateral Adrenal Non-Hodgkin Lenfomanın Neden Olduğu Semptomatik Adrenal Yetmezlik

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Abstract

Secondary involvement of the adrenal gland with non-Hodgkin's lymphoma has been reported to occur in up to 25% of patients during the course of the disease. A 45-year-old man presented with a several month history of nausea, fatigue and weight loss. His medical history was unremarkable. Abdominal computed tomography (CT) was performed and showed bilateral adrenal massive masses measuring 10x7.5 cm on the left and 4.8x4 cm on the right. He developed adrenal insufficiency in the follow-up period. The patient was started on replacement dose of prednisolone. A positron emission tomography-CT scan was acquired for further staging of the disease and showed intense fluorodeoxyglucose accumulation in both adrenal glands, additionally a slight fluorodeoxyglucose accumulation was observed in the ileocecal site. He did not accept adrenal biopsy or surgery. Histopathological examination of the ileocecal site revealed diffuse large B-cell lymphoma. He was administered rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone chemotherapy. Bilateral adrenal lymphoma is a rare entity when compared with the incidence of adenoma and adrenal metastases of other cancers. Adrenal insufficiency may be the primary symptom of presentation, especially with bilateral involvement as in bilateral adrenal lymphoma.
Keywords: Bilateral adrenal lymphoma, adrenal failure, positron emission tomography-computed tomography

Öz

Non-Hodgkin lenfoma seyrinde adrenal bez tutulumu hastaların %25'inde bildirilmiştir. Kırk beş yaşında erkek hasta bir kaç aydır olan bulantı, yorgunluk ve kilo kaybı nedeniyle başvurdu. Medikal hikayesinde özellik yoktu. Abdomen bilgisayarlı tomografide (BT) sağ adrenalde 4,8x4 cm ve sol adrenal 10x7,5 cm boyutlarında büyük kitleler tespit edildi. Takipte hastada adrenal yetmezlik gelişti. Hastaya prednizolon başlandı. Hastalığın yaygınlığının değerlendirilmesi için yapılan pozitron emisyon tomografisi-BT'de her iki adrenal bezde ve daha az yoğun olarak ileoçekal bölgede florodeoksiglikoz birikimi gözlendi. Hasta adrenal biyopsi veya cerrahi kabul etmedi. İleoçekal bölgeden biyopsi yapıldı. Histopatolojik incelemede diffüz büyük B-hücreli lenfoma olarak rapor edildi. Hastaya rituksimab, siklofosfamid, doksorubisin, vinkristin ve prednizon kemoterapisi uygulandı. Bilateral adrenal lenfoma, adrenal adenom ve adrenal metastaza göre daha nadir gözükmektedir. Adrenal yetmezlik özellikle bilateral adrenal tutulumun olduğu bilateral adrenal lenfoma olgularında ilk tanı anında karşımıza çıkabilir.
Anahtar kelimeler: Bilateral adrenal lenfoma, adrenal yetmezlik, pozitron emisyon tomografisi-bilgisayarlı tomografi

Introduction

The adrenal gland is a frequent site of neoplastic disease resulting in bilateral adrenal metastases, and lymphoma may occasionally involve the adrenal glands. Adrenal insufficiency is infrequently reported in patients with adrenal metastases. We present a case of a 45-year-old man with bilateral adrenal NHL presenting with adrenal insufficiency.

Case Report

A 45-year-old man with no prior medical history was admitted for a four-month history of abdominal pain, nausea, fatigue, and weight loss. Examination of head and neck was unremarkable; no abnormality was found in chest and abdomen; there was no evidence of lymphadenopathy or abnormal skin lesions. Routine laboratory investigations did not reveal any pathology. Ultrasonography of the abdomen demonstrated bilateral adrenal masses. Computed tomography (CT) scan of abdomen showed bilateral adrenal masses measuring 10x7.5 cm on the left and 4.8x4 cm on the right. Based on the CT findings, a provisional diagnosis of adrenal metastases or possible lymphomatous involvement of the adrenal was made.

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The hormonal profile including basal serum cortisol and adrenocorticotropic hormone (ACTH) levels were 9 µg/dL (5-25) and 225 pg/mL (0-46), respectively. During follow-up, laboratory examination showed hyponatremia with a sodium level of 127 mEq/L (136-147) and hyperpotasemia with a potassium level of 5.4 mEq/L (3.5-5.1). Rapid cosyntropin (analog of ACTH) stimulation test, using 250 µg of intravenous cosyntropin, demonstrated plasma cortisol concentrations at 0, 30, and 60 min of 8.0, 9.8, and 10.5 µg/dL, respectively, strongly suggesting adrenal insufficiency. The patient was started on replacement dose of prednisolone (7.5 mg/day). As the patient had bilateral adrenal masses we ruled out pheochromocytoma with normal urinary fractionated metanephrines. An 18-fluorodeoxyglucose (FDG) positron emission tomography (PET)-CT scan showed intense FDG accumulation in both adrenal glands, additionally a slight FDG accumulation was observed in the ileocecal region (Figure 1). Ileocecal biopsy was performed. Histopathological examination revealed diffuse large B-cell lymphoma, which were immunohistochemically positive for CD20, but negative for CD3. He was administered rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) chemotherapy. Due to persistent hyponatremia, fludrocortisone was added to prednisolon therapy at follow-up. An 18-FDG PET-CT scan performed after administration of four cycles of R-CHOP chemotherapy showed that previous adrenal masses had nearly disappeared (Figure 2). He died of respiratory insufficiency caused by aspiration pneumonia after six cycles of R-CHOP chemotherapy.

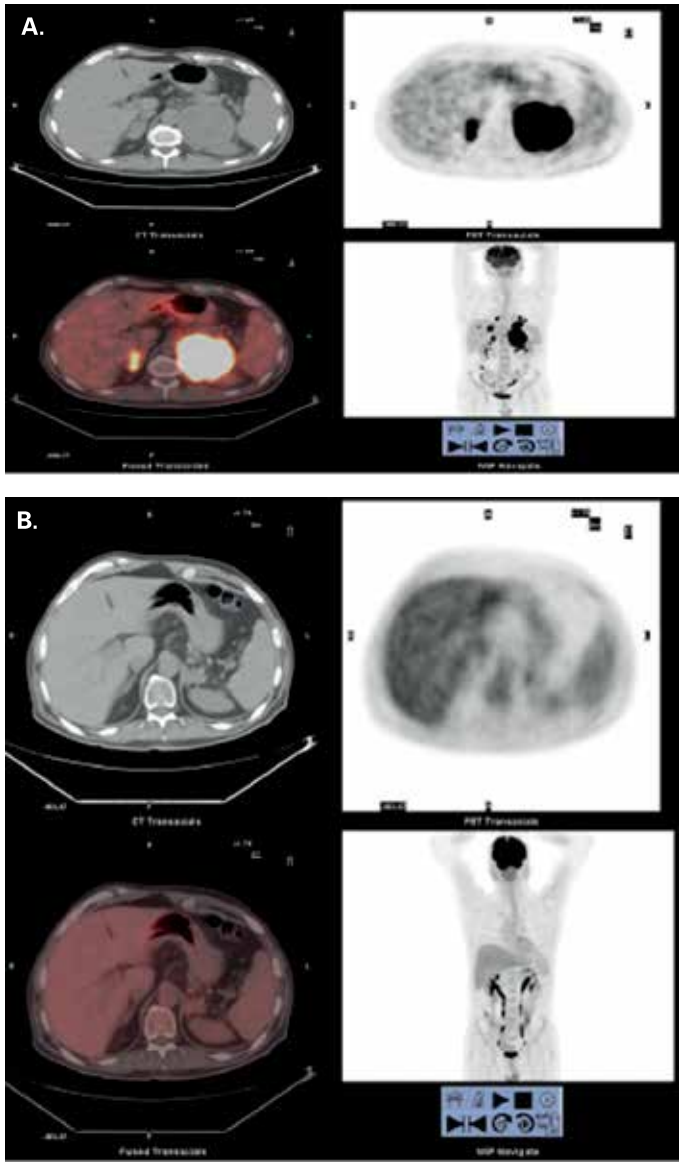

Figure 1. A and B positron emission tomography-computed tomography show focus of intense fluorodeoxyglucose uptake in bilateral adrenal masses. The left adrenal mass is bigger than the right. There is another focus with fluorodeoxyglucose uptake in ileocecal site

Discussion

Bilateral adrenal masses include a spectrum of disease: neoplastic disorders (metastases, lymphoma, bilateral phaeochromocytoma, adrenocortical carcinoma, and myelipoma); longstanding congenital adrenal hyperplasia and macronodular adrenal hyperplasia; infections such as tuberculosis and histoplasmosis; and adrenal heaemorrhage (such as catastrophic antiphospholipid syndrome) (1). Most bilateral adrenal masses are metastases from malignant tumors and adrenal insufficiency in patients with cancer is gradually due to bilateral adrenal destruction by metastases (2). Adrenal metastases occur late in the course of disseminated cancers and are common in autopsy findings (2). Lung, breast, gastric, renal, and colon carcinomas, melanomas and malignant lymphomas, and usually non-Hodgkin's lymphoma (NHL) spread to the adrenal glands. Adrenal metastases may not be identified easily since many of them are asymptomatic. Lymphoma may

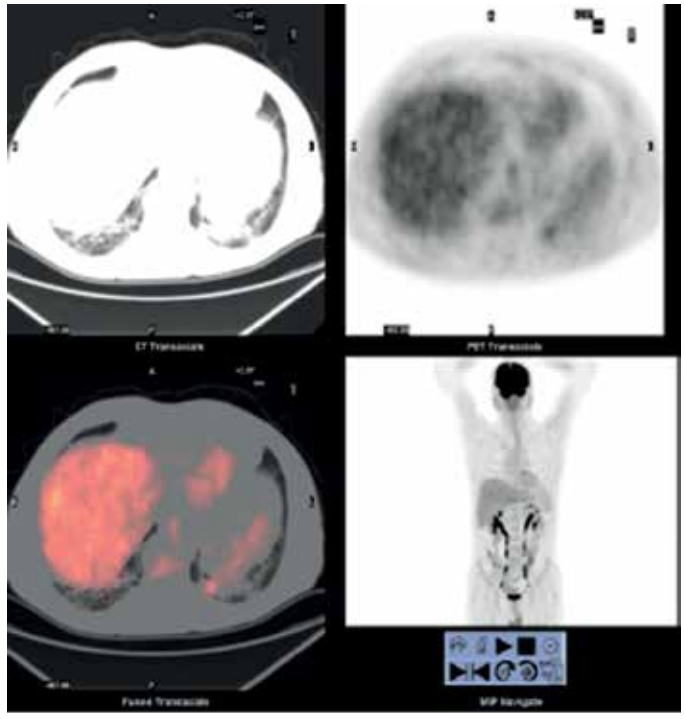

Figure 2. A follow up positron emission tomography scan shows significant reduced fluorodeoxyglucose uptake in bilateral adrenal masses

rarely involve the adrenal glands. More commonly, the adrenals are secondarily involved. The incidence of secondary adrenal involvement in NHL detectable on CT scan is about 5% (3) and increases to 25% when autopsy cases are included (4). Adrenal insufficiency may be the primary symptom of presentation, especially with bilateral enlargement as in bilateral adrenal lymphoma (5). Primary adrenal lymphoma is a rare disease with a mean survival period of thirteen months. The overall survival time depends on early diagnosis and treatment (2). In our case, as the patient did not accept adrenal biopsy, thus, further evaluation for adrenal metastasis, PET was performed which revealed a mass in the ileocecal region.

In conclusion, secondary bilateral adrenal involvement of NHL is often consistent with adrenal insufficiency. The present case indicates that adrenal lymphoma should be suspected in patients with bilateral adrenal masses or adrenal insufficiency.

Ethics

Informed Consent: It was taken.
Peer-review: Internal peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Şafak Akın, Concept: Şafak Akın, Serkan Akın, Design: Şafak Akın, Serkan Akın, Data Collection or Processing: Şafak Akın, Serkan Akın, Murat Tuncel, Neşe Ersöz Gülçelik, Analysis or Interpretation: Şafak Akın, Literature Search: Şafak Akın, Neşe Ersöz Gülçelik, Writing: Şafak Akın, Serkan Akın.
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