Primary bilateral non-Hodgkin’s lymphoma of the adrenal gland

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INTRODUCTION
In general, at least 90% tissue destruction is needed to affect the function of the adrenal gland; therefore, adrenal failure due to malignancies involving the adrenal gland is very rare. However, 61% of cases of primary adrenal lymphoma (PAL) have adrenal insufficiency, which may be due to the late detection of PAL because of the late onset of symptoms.

Despite the fact that secondary adrenal lymphoma occurs in 25% of nodal non-Hodgkin’s lymphoma (NHL) cases, primary lymphoma of the adrenal gland is rare. Bilateral PAL (B-PAL) accounts for 75% of PAL cases. Epstein–Barr virus positivity is observed in over 50% of such patients.

An incidentaloma (a mass lesion >1 cm in diameter, accidentally discovered by radiologic examination) is not common as a presenting problem in PAL, and its most frequent primary symptoms are B-symptoms (systemic symptoms of fever, night sweats, and weight loss which can be associated with both Hodgkin’s lymphoma and NHL), pain, and fatigue.

Male predominance is reported and the mean age is 62 years. The available literature shows that the median size of PAL is around 8 cm. The frequency of lymphadenopathy and bone marrow involvement at diagnosis are 18% and 6%, respectively. On ultrasound, PAL may present as hypoechoic heterogeneous masses. On computed tomography (CT) scan, PAL is usually described as low density with mild-to-moderate enhancement. Herein, we report the case of a young man with a high-grade lymphoma of the adrenal glands presenting as bilateral adrenal masses.

CASE REPORT

A 38-year-old male was referred to our clinic with symptoms of abdominal pain, nausea, vomiting, and fever for a month. During this period, the patient’s weight had reduced by 10 kg. An ultrasound study was requested reporting a bilateral mass corresponding to the adrenal region. In the next step, an abdominopelvic CT scan with and without oral and intravenous contrast was performed, which revealed...
two adrenal masses of 115 mm × 66 mm and 65 mm × 43 mm in the right and left adrenal glands, respectively. There was no retroperitoneal lymphadenopathy [Figure 1].

At the time of admission, primary physical examination and basic laboratory tests were normal. Blood pressure and temperature were within normal limits and no peripheral lymphadenopathy was detected. On physical examination, the thyroid gland was normal on palpation and no skin hyperpigmentation was seen. Laboratory tests revealed the following results: white blood cell (WBC) count = 7.5 × 10^9/L, hemoglobin (Hb) level = 10.8 g/dL, platelet (PLT) count = 179 × 10^9/L, erythrocyte sedimentation rate (ESR) = 6 mm/h, serum creatinine = 1.2 mg/dL, serum sodium = 144 mEq/L, and serum potassium = 4.7 mEq/L; a urine analysis was normal.

Around 10 days after the initial tests, additional tests were performed for the patient; their results were as follows: WBC = 2.4 10^9/L, Hb = 7.6 g/dL, PLT = 60 × 10^9/L, serum sodium = 125 mEq/L, and potassium = 5.5 mEq/L. Cortisol, metanephrine, normetanephrine, and vanillylmandelic acid of the 24-h urine output were 39 µg, 46 µg, 60 µg, and 1.6 µg, respectively. Morning serum cortisol was normal (13.5 µg/dL) whereas the adrenocorticotropic hormone level was elevated (90 pg/mL).

Furthermore, lactate dehydrogenase level was 893 U/L and ESR had risen to 63 mm/h, which demonstrated the rapid progression of the disease and indicated the onset of adrenal insufficiency. A biopsy sample was taken from the aforementioned masses under the guide of CT scan. This CT scan, done 20 days after the first CT scan showed rapid progression of the disease with further enlargement of the adrenal masses along with retroperitoneal lymphadenopathy [Figure 2].

The Pathological examination of the biopsy reported high-grade B-cell lymphoma with immunohistochemistry positive for CD20 and negative for CK and CD3. Therefore, we decided to perform bone marrow biopsy and chest CT scan. The bone marrow biopsy was negative for malignancy, but the chest CT scan revealed a few lymph nodes. During this evaluation study, the patient's condition deteriorated significantly, so after the pathological diagnosis, chemotherapy was immediately initiated for the patient by an oncologist; the patient was treated with six cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone (R-CHOP) chemotherapy. The CT scan performed 2 months after the end of R-CHOP chemotherapy showed regression of the bilateral adrenal mass [Figure 3], and subsequently, the patient’s general condition improved significantly.

**DISCUSSION**

Primary bilateral NHL of the adrenal gland has to be considered whenever bilateral adrenal masses are present on CT scan images. PAL usually has no excretory endocrine function and the symptoms are due to the pressure effect of the mass,[2] whereas adrenal insufficiency usually exists.
Nevertheless, a few articles have reported normal adrenal function in B-PAL,\(^4\) similar to our patient which seems to be the reason for the early diagnosis in such cases.

In the present case, CT-guided biopsy was done for bilateral adrenal masses which appeared well circumscribed and homogeneous with low enhancement on CT scan and had normal adrenal function. The diagnosis of primary bilateral NHL was confirmed, in view of no lymphadenopathy on physical examination and initial imaging, no evidence of other malignancies, and a negative bone marrow biopsy for malignancy, along with the pathology report of adrenal biopsy reported as large B-cell lymphoma.

As surgery is not recommended in these patients, we referred the patient to an oncologist for further chemotherapy and follow-up. Although the prognosis of PAL is very poor and complete remission with chemotherapy has been reported only in a few articles,\(^6\) 6 months after diagnosis and 2 months after chemotherapy termination, this patient was progressing well.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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