Case Report

An uncommon unilateral primary T cell adrenal lymphoma: A case report

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ABSTRACT

Primary adrenal lymphoma (PAL) is a rare disease with rapid progression. We present a case of PAL with T cell lymphoma in an 81-year-old male who initially presented with altered mental status and extreme weight loss. It is important to consider PAL in patients found to have adrenal masses to allow for prompt diagnosis and initiation of treatment.

1. Introduction

Primary adrenal lymphoma (PAL) is an aggressive lymphoma with approximately 250 cases reported [1]. Patients can present with malaise, B symptoms, as well as rapid weight loss. PAL should be included in the differential diagnosis if imaging shows unilateral or bilateral adrenal masses. Pathology-directed chemotherapy is the first line treatment modality.

2. Case presentation

An 81-year-old male with a medical history of benign prostatic hyperplasia presented with a one day history of altered mental status (AMS), hypoxia and a reported 80 pound weight loss over four months. Patient’s only past medical history was benign prostatic hyperplasia status post transurethral resection of the prostate (TURP). He was on no medications at home. Family medical history was unknown.

At presentation, vital signs were notable for a heart rate of 105. Physical exam findings were pertinent for disorientation to time. Neurologic exam was otherwise within normal limits. CBC (complete blood count) was significant for white blood cell (WBC) count of 3.0 K/mm³, hemoglobin of 13.6 gm/dL and platelets of 114 K/mm³. Comprehensive metabolic panel (CMP) was significant for mild hypokalemia with Na of 134 mmol/L, elevated total bilirubin of 1.2 mg/dL and AST of 101 Units/L. Thyroid stimulating hormone, B12 and ammonia were within normal limits. Syphilis serologies were negative. Computed tomography angiography (CTA) of chest was performed that showed no clonal B-cell or aberrant T-cell populations. He underwent CT-guided biopsy of the adrenal mass and the pathology was reported to be CD3+ T cell lymphoma. The patient rapidly declined and he died on day 7 of his hospitalization due to respiratory failure.

3. Discussion

Lymphoma infrequently involves the adrenal glands, with non-Hodgkin’s lymphoma being more common than Hodgkin’s disease at this site. Differential diagnoses include adrenocortical carcinoma, pheochromocytoma, metastatic disease or cortical adenoma. PAL is a rare and aggressive malignancy with approximately 250 cases reported [1]. PAL accounts for less than 1% of non-Hodgkin lymphomas with diffuse large B cell lymphoma accounting for 78% of PAL [2,3]. T cell lymphoma accounts for approximately 7% of PAL cases [4]. PAL typically occurs in men aged 60–70 years. Patients may initially present with abdominal pain, B symptoms including fever and weight loss, adrenal insufficiency (AI), and elevated LDH [4,5]. AI develops in about 60% of PAL patients, with increased risk in those with bilateral adrenal involvement, older age and history of autoimmune disease [3]. One invasive, malignant mass involving the left adrenal gland and upper pole of the left kidney measuring 9.9 × 7.0 × 9.5 cm (Fig. 1). Brain magnetic resonance imaging (MRI) was ordered to investigate for primary central nervous system lymphoma and showed no acute abnormalities.

During his admission, the patient developed a bacterial pneumonia which was treated with vancomycin and piperacillin/tazobactam. During his hospital course he developed progressively worsening mental status. The patient became less verbal and had poor oral intake. Further labs revealed lactate dehydrogenase (LDH) elevated at 1041 Units/L and worsening pancytopenia. Flow cytometry was performed that showed no clonal B-cell or aberrant T-cell populations. He underwent CT-guided biopsy of the adrenal mass and the pathology was reported to be CD3+, CD30+ T cell lymphoma.

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A study showed bilateral adrenal involvement in 60% of cases [6]. Our patient was an elderly male without any history of autoimmune disease. He did not develop any signs or symptoms of AI with labs showing only minimal hyponatremia, this may have been due to only unilateral adrenal involvement rather than bilateral, as well as absence of autoimmune diseases. It should be considered that with prolonged disease course, AI symptoms may have developed in our patient with disease progression. The pathophysiology of PAL remains unclear with one hypothesis being that PAL develops from hematopoietic tissue similar to adrenal myelolipoma [7]. The lesions are typically initially noted on MRI or CT imaging. Fluorodeoxyglucose positron emission tomography (FDG PET) can be used to differentiate benign and malignant lesions as well as show involvement at other sites [8]. Diagnosis also requires no prior history of lymphoma and if there is extra-adrenal involvement the dominant lesion should involve the adrenal gland [9]. Histopathology is performed for definitive diagnosis with samples from biopsy, surgical excision, or autopsy. Our patient’s pathology was identified as CD3+ , CD30+ T cell lymphoma. T cell adrenal lymphoma is rare with a poor prognosis with nearly half of PAL patients reported in the literature passing away within 1 year [3]. Treatment modalities include surgery, chemotherapy, radiation, as well as corticosteroid replacement in those with AI [10]. A study with 31 patients diagnosed with diffuse large B cell PAL demonstrated a remission rate of 55% and 2 year survival rate of 68% with cyclophosphamide, doxorubicin, vincristine and prednisolone (CHOP) therapy [11]. Systematic review by Rashidi et al. of 149 patients had a 12 month survival rate of only 20%, however it should be noted that only 21 of these patients received CHOP therapy with 106 patients receiving another form of chemotherapy [3]. One study found that rituximab in addition to CHOP therapy had higher response rates as well as higher overall survival times compared to CHOP therapy alone [12]. There is little data regarding efficacy of these treatments specifically for T cell PAL. Our patient presented late in the disease process and had a significant functional decline which would have precluded chemotherapy as a treatment option.

4. Conclusion

This case highlights the importance of including a rare subtype of PAL in the differential diagnosis of adrenal masses so that prompt diagnosis can be made for better clinical outcome of this aggressive and life-threatening illness. Diagnosis should be made by obtaining a biopsy. Treatment options include chemotherapy, radiation and surgery. Our patient passed away before any pathology directed chemotherapy could be initiated.

Ethical approval

This study was approved by Ethics Committee.

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Author contribution statement

Kiranpreet Gosal: Conceptualization, Writing - original draft, Writing - review & editing. Mitsy Barr: Conceptualization, Writing - original draft, Writing - review & editing, Data curation. Pooja Patel: Data curation, Conceptualization, Writing - review & editing. David Moccia: Writing - review & editing.

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Guarantor

Kiranpreet Gosal, D.O.

Consent

Written informed consent was not obtained from the patient. The head of our medical team has taken responsibility that exhaustive attempts have been made to contact the family and that the paper has been sufficiently anonymized not to cause harm to the patient or their family. A copy of a signed document stating this is available for review by the Editor-in-Chief of this journal on request.

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Declaration of competing interest

The authors declare no conflict of interest.

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Fig. 1. CT imaging showing left adrenal and left upper kidney pole mass measuring 9.9 × 7.0 × 9.5 cm.
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