

Primary Adrenal Lymphoma

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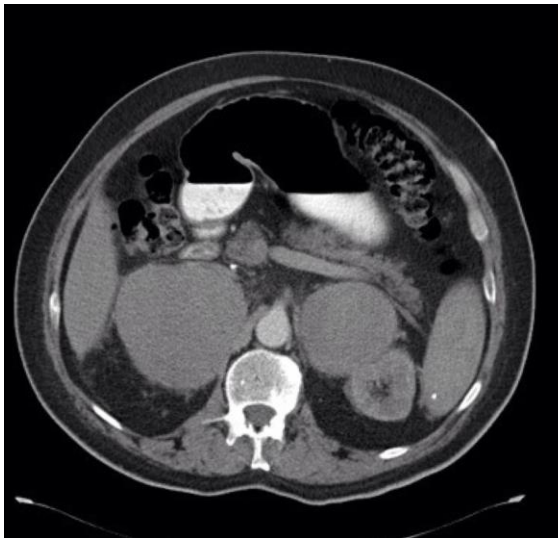


Fig. 1 Baseline CT Abdomen

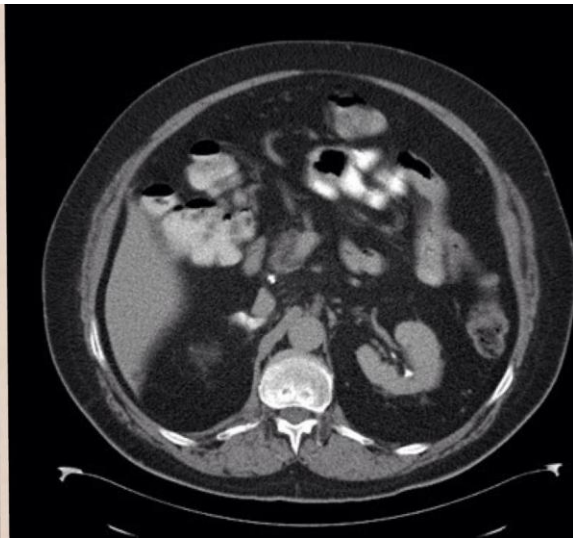


Fig.2 CT Abdomen, 2 year follow-up

A 77 year-old Caucasian male complained about a 3-month history of progressive weakness, intermittent fevers and a 40 pound weight loss. He presented to the ED with abdominal pain, a hemoglobin level of 8.3 g/dl, MCV 95 fl. The patient had a negative gastrointestinal evaluation for blood loss sources completed a month before. LDH level was elevated at 669 U/L, beta-2 microglobulin at 4.3 mg/L, electrolytes, liver enzymes, thyroid studies and renal function tests read normal. Abdominal CT scan revealed a 10 x 10 x 9.5 cm mass on the right adrenal gland and a 10 x 7.6 x 7 cm mass on the left side. There was no adenopathy in the para-aortic or mesenteric area (Figure 1). CT guided biopsy showed CD45+ and CD20+ large cells with high nuclear:cytoplasm ratio, as well as scant cytoplasm consistent with diffuse large B-cell lymphoma (DLBCL). Due to a high apoptotic rate, further studies were done, MYC/IGH gene rearrangement was performed with negative results. Bone marrow biopsy did not show lymphomatous involvement. Staging PET-CT scan showed bilateral adrenal masses with a SUV as high as 27. There was no extra-adrenal uptake.

With the diagnosis of stage IIEB, primary adrenal DLBCL, R-IP1 2, the patient received 6 cycles of R-CHOP achieving complete remission (Figure 2). He continues to be in remission 4 years after diagnosis.

Primary adrenal non-Hodgkin's lymphoma (PAL) is an extremely rare neoplastic disease, usually presenting with bilateral large adrenal masses, sometimes accompanied by adrenal insufficiency or hypercalcemia. The male-to-female ratio is 2.2:1; most frequently seen in older men with a mean age at diagnosis of 68 years [1]. The etiology is still unknown but associated with some autoimmune diseases [2] [3] [4] such as autoimmune adrenalitis with previous adrenal insufficiency [5] [6] and autoimmune hemolytic anemia[7]. The most common histologic subtype is diffuse large B-cell lymphoma with a non-germinal center B-cell phenotype and frequent BCL6 gene rearrangement [8]. It has a very poor prognosis with a 1-year survival rate lower than 20% [9]. Because of its rare occurrence, there are no clear treatment protocols. It is usually treated with chemotherapy with or without radiotherapy [2]. Surgery is not recommended. [1]



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