Primary Bilateral Arenal Lmphoma: A case report

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Primary adrenal lymphoma is extremely rare. We report a case with primary bilateral adrenal lymphoma in a young male patient. He presented with abdominal pain and weight loss. Pathologic study revealed malignant lymphoma, diffuse large cell type (T- cell origin). Patient recieved combination chemotherapy and radiation therapy but 22 months after diagnosis died because of progression of disease.
Key words: adrenal gland; primary lymphoma; clinicopathologic features.

Introduction

The adrenal gland is involved in approximately 25 % of patients with diffuse malignant lymphoma.[1] In contrast primary adrenal lymphoma is extremely rare and about 75 cases have been reported world wide[1,4] and bilateral adrenal lymphoma is more rare than unilateral.[1-10,14-20] Because of its rarity we report here a young man with primary bilateral adrenal lymphoma and describe his clinical manifestations, histologic features, diagnosis, management and clinical course.

Case report

A 20 - year - old man from Iran admited to us with abdominal pain and weight loss (12 kg in 3 months). His pastmedical history was negative. Vital signs and examination of chest were normal. There was no lymphadenopathy and hepatosplenomegaly but bilateral massive masses were found in his flanks. Labratory tests and chest x-ray were normal. Abdominal ultrasonography and CT scan revealed bilateral massive adrenal masses (fig 1&2).

CT guided adrenal fine needle aspiration biopsy was performed . It revealed malignant lymphoma , diffuse large cell type with T-cell phenotype based on immunohistochemistry study [LCA (CD45) and CD3 were positive ; CD20 , NSE , cytokeratin and EMA were negative] (fig 3).

Bone marrow aspiration and terphine biopsy were normal. Adrenal function test was normal. Combination chemotherapy was intiated with cyclophosphamide 750 mg/m2, Adriamycine 45 mg/m2, vincristin 2mg and prednisolon. This treatment was repeated every 21 days for 6 cycles.

After completion of this treatment abdominal CT scan was performed and revealed complete regression of abdominal masses (fig 4&5).

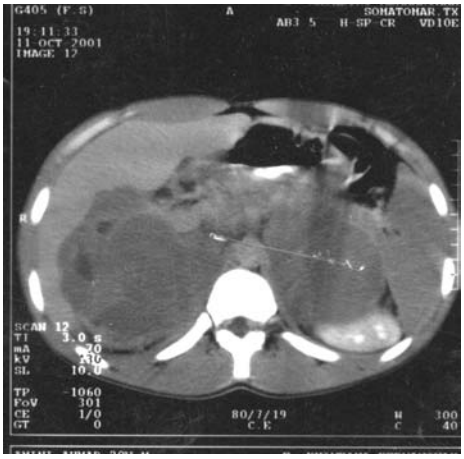


Figure 1

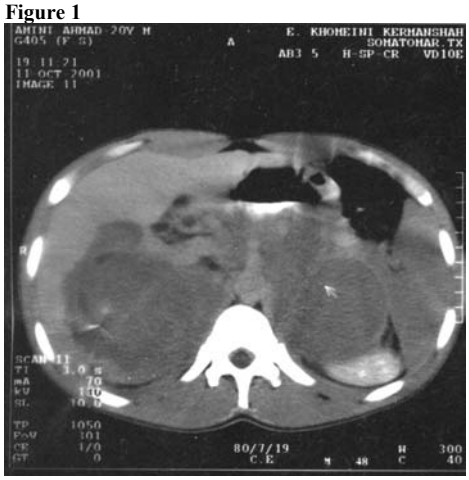
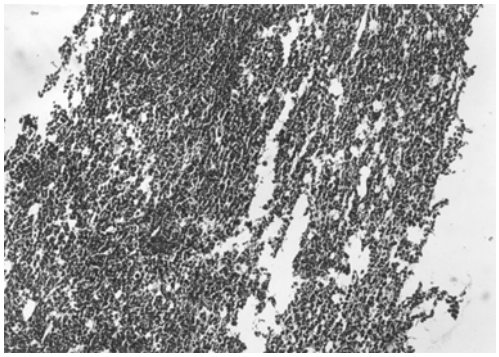
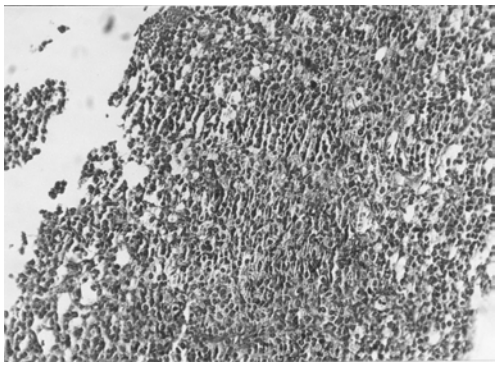


Figure 2

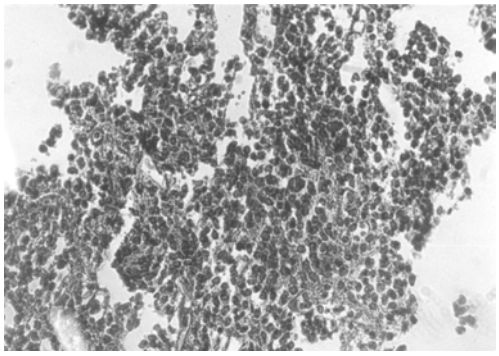
fig 1&2 : Abdominal CT Scan revealed bilateral adrenal masses in patient with abdominal pain and weight loss . (before first hemotherapy)



3 a



3 b



3 c

Figure 3:a:lymphoma cells infiltrating adrenal in a diffuse pattern(x100) b: Higher magnification showing largecell lymphoma . No adrenal tissue is identified (x 400) . c: Oil immersion view , revealing large lymphoma cells (x 1000).

The patient was under observe. 10 months later cameback because of abdominal pain. Abdominal CT scan was performed and unfortunately showed relapse of adrenal masses.

MINE protocol, Etoposide, Ifosfamide and Mitoxantrone, began for 6 cycles every 3 weeks. Abdominal CT scan was performed after this protocol, but the masses were still present. The patient was refferred for radiation therapy, but there was no good response to this treatment and patient died 4 months later because of disease progression, and overwhelming infection.

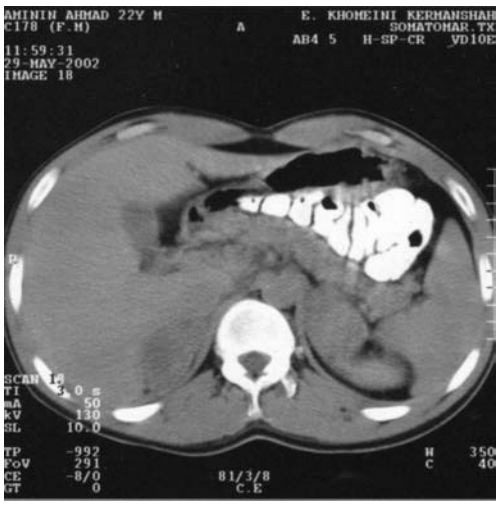


Figure 4

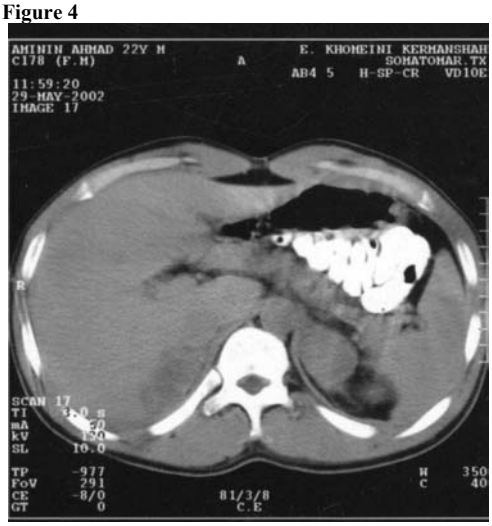


Figure 5

Figure 4 and 5: Abdominal CT Scan after first chemotherapy in the same patient.

Discussion

It suggested that primary adrenal lymphoma is a distinct entity and should be considered in patient with an adrenal mass, without lymphadenopathy or organomegaly, with or without Addison's disease and elevated serum LDH.[1, 15] Medical imaging (Ultrasonography, CT scan, MRI and Ga scintigraphy) is non specific and biopsy with pathologic examination remains the most reliable diagnostic method.[1, 2, 5, 13, 21] Most of these tumors have a high grade histology, almost with the B phenotype.[5, 7, 12, 22] Treatment modalities include surgery, combination chemotherapy, surgery followed by chemotherapy and/or radiation therapy.[4, 5, 7, 8, 10, 19, 24]

In this report our patient was a 20 year old man with primary bilateral adrenal lymphoma, who was the youngest between reported cases. He presented with abdominal pain, weight loss and flank masses but, in contrast to most of reported cases with bilateral adrenal lymphoma, hasn't

any symptoms or signs of adrenal insufficiency.[5, 7, 14, 15, 23]

Diagnosis of disease in our patient, such as most of other reported cases, was based on CT guided needle biopsy with diffuse large cell type but T- cell phenotype.

Our patient treated with chemotherapy, intially with good response, but eventually relapse and died because of tumor progression despite chemotherapy and radiation theraphy. He survived 22 months after diagnosis of disease.

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