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Primitive Bilateral Adrenal Lymphoma Phenotype T: A Case Report And Review of Literature

Soumeya Fedala[1], Ali el mahdi Haddam[2],Radhia si Youcef[3],
Bab el oued Hospital, Department of Endocrinology,
Department of diabetology Bab el oued hospital,
Bologhine hospital, Department of Biochemistry.

Abstract: *Lymphoma adrenal is a rare cause of adrenal tumor(0,5%). Bilateral primary lymphoma adrenal phenotypeT is exceptional. We report observation of a patient 56 years old hospitalized for exploration and therapeutic management of two large adrenal masses discovered on CT imaging after back pain, and weight loss. Physical examination revealed a patient asthenic, with no signs of hypersecretion. The rest of the examination was unremarkable and research call signs primary neoplasm was negative. A hormonal balance showed a low cortisol with a hight ACTH level.This which required start hormone replacement therapy to hydrocortisone.The magnetic resonance imaging objectivied large masses without signs of infiltration. There were no signs of locoregional infiltration and no deep lymph nodes. The patient is operated by addition midline laparotomy bypassing the umbilicus to the left and extending by 4cm below, with removal of the left adrenal tumor. At the right, the tumor mass cannot be eradicated. It is simply biopsied. Histological examination completed by immunohistochemical study concluded to an anaplastic large cell lymphoma, phenotype T with intense membrane staining and diffuse with CD3 and a lack immunostaining with CD20 and CD 30.Additional is undertaken at home postoperatively according CHOP. Unfortunately, the patient died at the waning of his fifth treatment in an array of septic shock.*

Keywords*: Adrenal masses;Bilateral primary lymphoma adrenal; phenotype T; chemotherapy*

I. Introduction

Primary lymphoma is an uncommon cause of adrenal tumor, found in 0.5% of cases[1]. A predominance of diffuse large B cells is reported in most publications. Bilateral involvement of T phenotype is exceptional and has been observed in six cases. Characterized by a very difficult diagnosis and prognosis, it must be suspect in the presence of bilateral adrenal masses with rapidly increasing volume. We report about this a new observation.

Observation:

Mr. O. S. 31 years old, married, had since two months a bilateral low back pain associated to a poor general condition, asthenia and weight loss of 5kg. An ultrasound is then performed showing large adrenal masses. The patient is admitted in the endocrinology department for further exploration.The examination at the admission found an asthenic patient with a body mass index of 23 kg/m2 and a blood pressure at 110/60mmHg. Physical examination revealed no skin pigmentation or other signs of adrenal dysfunction. It is not found palpable masses, hepatomegaly nor splenomegaly. There was also no lymphadenopathy or signs of call for a primary neoplasm or infiltrative disease.In paraclinical plan, the non specific biological exploration objectified a lower serum sodium limit to 136 mEq / l and a moderate inflammatory syndrome (TABLE I).

Table I: Results of non-specific balance

parameters	Result	Norms
Glycemia(g/l)	0,65	0,70-1
Serum electrolytes natremia (meq/l)	136	135-145
potassium(meq/l)	3,8	3,5-5
Sedimentation rate 1st hour	20	
2nd time	45	
serum albumin(g/l)	28,4	36-48
al globulins (%)	18	7-11
a 2 globulins(%)	22	13-19
NFS	GR : 4,82 10 6/mm3 ; GB : 4,4 10 3/mm3 ;Plaquettes : 285 10	
liver function tests alkaline phosphatase Total bilirubin, conjugated,	100	98-279
free (mg/l)		<10 ; < 2 ; < 8
Transaminases ASAT(UI/l)	6 ; 0,2 ;2	<41
ALAT (Ul/l)	75	<41
	110	

Hormonal exploration revealed the existence of a primary adrenal insufficiency which required the prompt start of hormone replacement therapy with hydrocortisone. The remaining balance was unremarkable (Table II).

Table II: hormonal results

parameters	Result	Norms
Urinary methoxylated derivatives	0,59	<1 mg/24h
blood Cortisol	111,24	154-638 (nmol/l)
ACTH	88	7,6-66,1(pg/ml)
SDHEA	16,43	133-441(ug/dl)
17OHP	5,67	1,5-7,2(nmol/l)
plasma oestradiol	16	0-56(pg/ml
plasma testosterone	12,21	10,41-41,64nmo/l
plasma FSH	1,6	0,7-11,1 MU/ml
plasma LH	1,1	0,8-7,6 MU/ml

The thoraco-abdominal CT scan showed two large heterogeneous processes tissue density with clear limits, measuring 77/41 mm right and 75/43 mm on the left. In magnetic resonance, these masses were isointense in T1, T2 hyperintense and enhanced heterogeneously low and after injection of contrast. There were no signs of locoregional infiltration and no deep lymph nodes (Fig. 1). A morphological staging (chest radiography, CT scan, bone scan, bronchoscopy gastroesophageal) looking for other tumor locations proved without anomalies.The patient is operated by addition midline laparotomy bypassing the umbilicus to the left and extending by 4cm below, with removal of the left adrenal tumor . At the right, the tumor mass was badly limited measuring 20x15cm about major axis and extending to the diaphragm and back-aortocaval without individualization of cleavage plane. Cannot be eradicated, the mass is simply biopsied. No profound lymphadenopathy was found. Histological examination completed by immunohistochemical study concluded to an anaplastic large cell lymphoma, phenotype T with intense membrane staining and diffuse with CD3 and a lack immunostaining with CD20 and CD 30(fig.2 and 3). Additional chemotherapy is undertaken at home postoperatively according CHOP (cyclophosphamide 750 mg/m2 per day , vincristine 2 mg / cycle, doxorubicin 50 mg / m 2 per cycle, prednisone 60 mg / day) . Cycles were repeated every 21 days. Unfortunately, the patient died at the waning of his fifth treatment in an array of septic shock.

II. Discussion

The adrenal NHL is very rare. It infiltrates secondary adrenal glands in 25% of cases, while the primitive adrenal involvement is very little reported. Less than 100 cases described in the literature[1] 75% of bilateral forms with a size greater than 5 cm[2]. A prédominance of diffuse large B cells was noted in most publications as the type T is exceptional. Men are more frequently affected than women with a peak incidence at the age of 62 years[1].The clinical signs are not specific. The diagnosis can be revealed by unexplained fever, abdominal pain or weight loss. Adrenal insufficiency is observed in approximately 86% of cases with evidence of an almost complete invasion of the adrenal gland.The lack of skin pigmentation in our patient despite the low rate of cortisol can be explained by the speed installation of adrenal insufficiency in our case. This adrenal insufficiency is more common in lymphoma type T (86% of cases: six out of seven) than in the lymphoma type B (56%: 27 of 48).[3] Asymptomatic in most cases, it should be sought systematically.Radiologically, abdominal ultrasound often shows a round mass, hypoechoic, with liquidiennes areas corresponding to hemorrhagic or necrotic areas.[4] The scanner usually evokes the diagnosis by showing a unilateral or bilateral adrenal large mass, heterogeneous without calcification or irregular or increased adrenal size.[4] Tumor mass is slightly enhanced by the radiological contrast product. Magnetic resonance of the adrenal mass has a hypointense signal on T1, with heterogeneous hypointense areas and heterogeneous hyperintense on T2.[2]The discovery of bilateral adrenal masses should be discussed in order of frequency: adrenal metastasis representing the 4th metastatic site after the lung, liver and bone; tuberculosis; malignant adrenocortical carcinoma; bilateral pheochromocytoma and some rare benign infiltration such as cryptococcosis, blastomycosis, histoplasmosis.[5] PET SCAN could be useful for differentiating benign from malignant lesion. It has a sensitivity of 92 to 100 % and a specificity of 80 to 100 %. [6,7] The main differential diagnosis arises with metastatic secondary injury. The radiological features are not specific, only histological study after laparotomy or fine needle biopsy guided by ultrasound or scanner used to make a diagnosis. To differentiate the primary or secondary nature of adrenal lymphoma, a staging should be performed. It includes a search for lymphadenopathy, splenomegaly, a chest radiography, a thoraco-abdominal CT scan, bone marrow biopsy and upper gastrointestinal endoscopy with biopsies. Only the negativity of these tests allows confirming the primitive character of lymphoma. The staging was negative in our patient.A reference assay of LDH and beta -2microglobulin will be made before treatment and a second one for the therapeutic monitoring. Some authors use the 18F-FDG PET Scan for postoperative monitoring.[8] Chemotherapy is the treatment of choice, however, surgery is indicated in rare cases, when the biopsy does not allow the diagnosis, in the case of tumor complicated by bleeding or to reduce the tumor mass prior to chemotherapy.[9,10]

Complète remission can be achieved after chemotherapy alone or combined with surgery, the duration of which varies from two months to one year.[3,11] Cases survival of eight years have been reported after combined treatment with surgery and radiotherapy without chemotherapy.[3] The prognosis of adrenal lymphoma and particularly lymphoma type T remains severe, with an average survival of 15 months. Death may occur in the acute period by adrenal insufficiency even before exploring the sick.[4]

III. Conclusion

Adrenal primary lymphoma is a very rare disease. Its diagnosis should be considered in the etiology of adrenal tumors. Radiological data are not specific, only the histological study allows a certain diagnosis. Chemotherapy is the treatment of choice although the prognosis remains poor in most cases.

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Figures

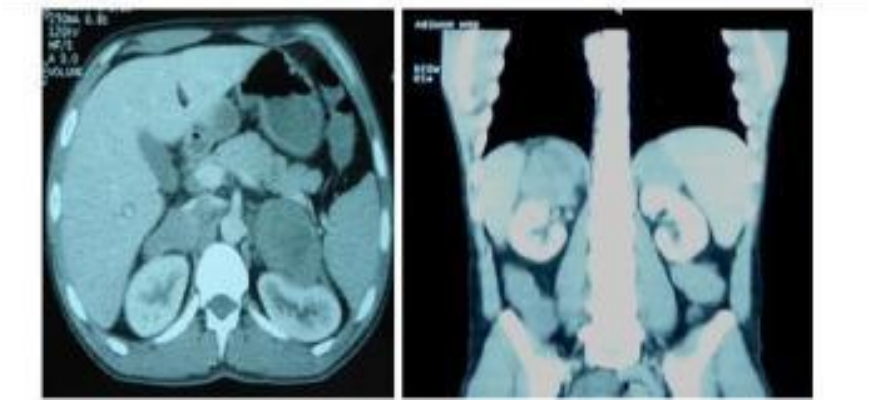
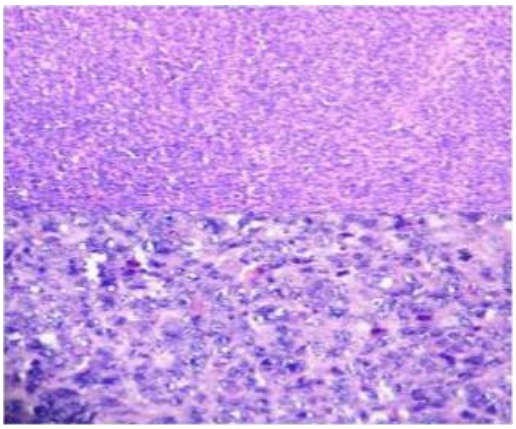


Fig 1: A+B :Abdsmiral CT.Axial section with cororal reconstruction image of fccth adranal tu mors



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