A RARE CAUSE OF UNEXPECTED BILATERAL ADRENAL GLAND ABNORMALITY

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INTRODUCTION

Bilateral adrenal gland enlargement can be caused by a number of endocrine, infective, inflammatory and neoplastic processes. We describe a rare haematological cause with multisystem involvement in an elderly woman.

CASE STUDY

A 79 yr old female with a background of hypertension and tablet controlled Type 2 Diabetes Mellitus presented to hospital with a four month history of progressive leg weakness, back pain and falls.

Examination revealed marked loss of power (2/5) in upper and lower limbs with foot drop.

Blood tests showed a normocyctic anaemia with raised ESR. A small paraprotein band was detected. Kidney, liver and bone profiles were unremarkable.

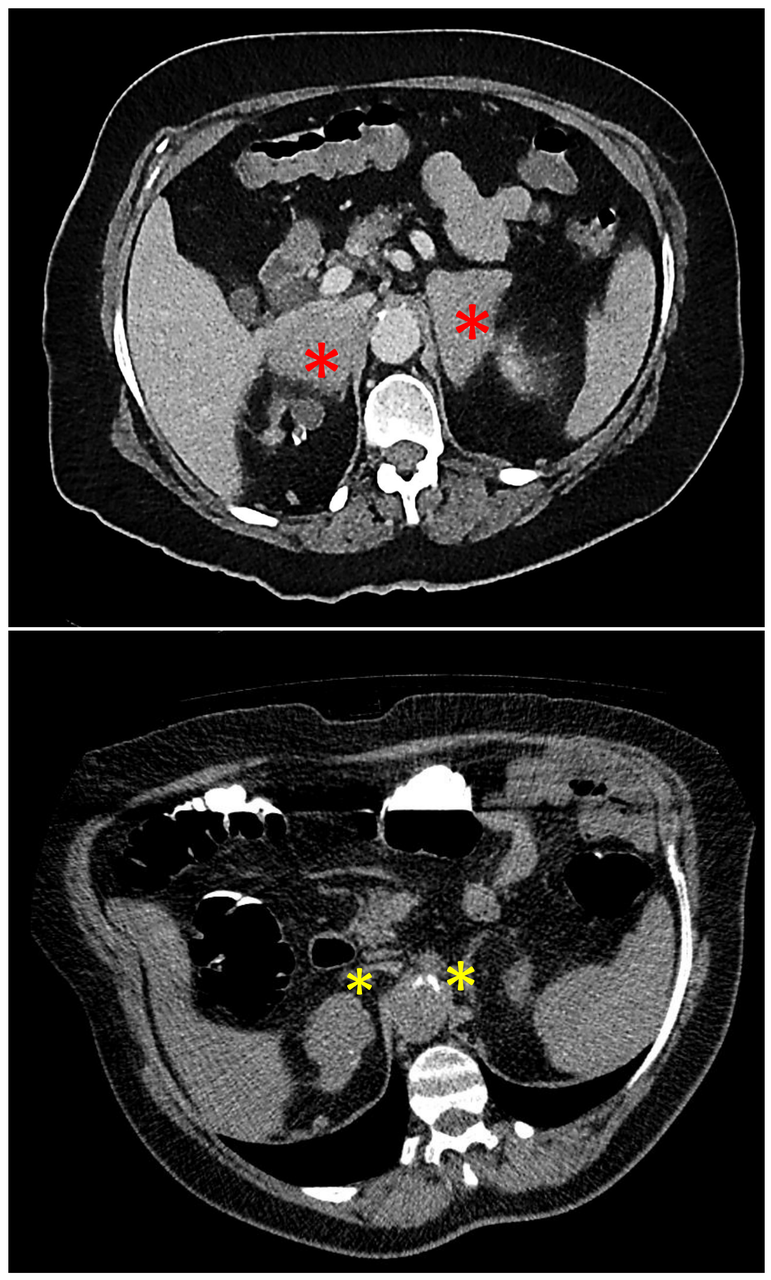
An MRI of spine showed degenerative change which did not explain the severity or pattern of weakness. Serial nerve conductive studies showed a rapidly progressive atypical large fibre sensori-motor axonal loss most in keeping with a paraneoplastic or vasculitic process. Subsequent negative autoantibody screen and sural nerve biopsy was not in keeping with the latter.

The patient underwent a CT of chest to pelvis which showed strikingly enlarged, smooth contoured adrenal glands of approximately 5cm diameter which had been normal in appearance four years prior.

Endocrine evaluation revealed normal 24h urinary (255 nmol/24h) and appropriate serum cortisol for acute illness (665nmol/L) with ACTH 7nmol/L. Plasma metanephrines and aldosterone were not elevated. There were no clinical features supporting androgen or cortisol excess.

Biopsy of the right adrenal gland showed replacement with malignant cells with an expression pattern typical of diffuse large B cell lymphoma with Ki-67 of 85%.

The patient was treated with high dose steroids but unfortunately, after a short trial of chemotherapy, died 6 weeks later.



Adrenal appearance at presentation (top) and four years previous (below)

DISCUSSION

Primary adrenal lymphoma (PAL) is extremely rare with fewer than 200 documented cases in the literature. Most cases (>70%) are bilateral and of the diffuse large B cell subtype, with adrenal insufficiency being a common presenting feature.

A review in 2008 by Ozimek et al (1) identified pointers towards a diagnosis of PAL as: bilateral adrenal enlargement >6cm; associated adrenal insufficiency; no systemic malignancy; age >60; male gender and short history, and thus this diagnosis should be considered in cases of primary adrenal failure in older patients, and when evidence of autoimmunity is lacking.

Diagnosis is made by adrenal biopsy and treatment is with combination chemotherapy (R-CHOP regime). Survival at 2 years is 68% in patients able to complete chemotherapy, but unfortunately this is an aggressive cancer and overall one year survival is only 20% (2) .

1. Ozimek A, Diebold J, Linke R, Heyn J, Hallfeldt K, Mussack T. Bilateral primary adrenal non-Hodgkin's lymphoma and primary adrenocortical carcinoma--review of the literature preoperative differentiation of adrenal tumors. Endocrine journal. 2008;55(4):625-38.

2. Rashidi A, Fisher SI. Primary adrenal lymphoma: a systematic review. Annals of hematology. 2013;92(12):1583-93.