

PRIMARY ADRENAL LYMPHOMA

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Case Summary

A 68 year old man diagnosed with a pulmonary embolus was found to have bilateral adrenal masses (image 1). Whilst awaiting outpatient follow up he was readmitted with pyrexia and fatigue. Repeat CT revealed the adrenal masses had grown significantly, the left in particular (image 2). There was biochemical evidence of adrenal insufficiency. CT guided adrenal biopsy elucidated the diagnosis of a high grade B cell lymphoma and R-CHOP chemotherapy was instituted. There has been an excellent response to treatment with rapid reduction in tumour size (image 3).

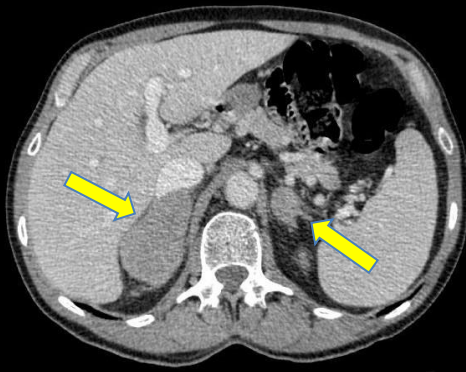


Image 1:

Initial CT following presentation with PE. Incidental finding of adrenal masses as indicated by the arrows.



Image 2:

Re-presentation with adrenal insufficiency. Increased size of adrenal masses demonstrated.

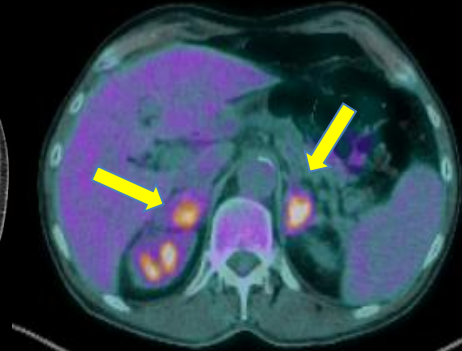


Image 3:

PET CT after initiation of R-CHOP. The metabolically active adrenal masses are significantly smaller.

PRIMARY ADRENAL LYMPHOMA AT A GLANCE

Diagnosis: lymphoma confirmed on histology +

- one or both adrenal glands involved; these are the primary lesions
- no previous history of lymphoma involving other organs

Prevalence: < 200 reported cases, accounts for <1% of all NHL

Gender: male > female

Age: 62 ± 14 years

Region: Asia (>50%), Europe (<30%)

Symptoms: pain, fatigue, pyrexia, weight loss, anorexia

Signs: adrenal insufficiency e.g. buccal/skin hyperpigmentation, may have hepatosplenomegaly or lymphadenopathy

Extra Laboratory tests: cortisol/synacthen test, LDH

Imaging modality of choice: PET CT, up to 70% are bilateral

Histological subtypes: 70% DLBCL

Treatment: chemotherapy dependent on histology e.g. R-CHOP for DLBCL, surgery, radiotherapy

Prognosis: insufficient data

Poor prognostic features:

- Advanced stage
- High LDH
- Bulky disease
- Older age at diagnosis

Thought to be associated with:

- Immunodeficiency
- EBV
- P53 and c-kit oncogene mutations

References:

Primary adrenal lymphoma: a systematic review. A Rashidi, S Fisher, Ann Hematol (2013) 92:1583–1593
Prognostic factors in primary diffuse large B-cell lymphoma of adrenal gland treated with rituximab-CHOP chemotherapy from the Consortium for Improving Survival of Lymphoma. Kim et al, Journal of Haem & Onc 2012 5;49