EP_A014

BILATERAL ADRENAL MASSES WITH RAPID DETERIORATION: A RARE CASE OF PRIMARY ADRENAL LYMPHOMA

Melody Tsen Shu Ling[1], Cheong Yee Weai[2], Hwee Ching Tee[2], Jin Hui Ho[2], Shireen Siow Leng Lui[2]
[1]Department of Medicine, Hospital Queen Elizabeth
[2]Diabetes & Endocrine Unit, Department of Medicine, Hospital Queen Elizabeth II

INTRODUCTION
Primary adrenal lymphoma (PAL) is a rare extranodal lymphoma, accounting for less than 1% of cases. It often presents with non-specific constitutional symptoms and features of adrenal insufficiency. The absence of pathognomonic findings frequently leads to delayed diagnosis, by which time the disease is often advanced, contributing to poor prognosis.

CASE
A 59-year-old male with type 2 diabetes mellitus and previously treated pulmonary tuberculosis presented with 1 month of lethargy, anorexia, weight loss, and abdominal pain. He was cachectic but hemodynamically stable (BP 120/81 mmHg; glucose 5.1 mmol/L), with otherwise unremarkable systemic examination. Laboratory evaluation revealed hyponatremia (120 mmol/L), hyperkalemia (6.0 mmol/L), and severe hypercalcemia (4.4 mmol/L). A short Synacthen test confirmed adrenal insufficiency.

Computed tomography demonstrated marked bilateral adrenal enlargement with bulky masses (8.9 × 8.7 × 8.7 cm right; 9.9 × 7.6 × 10.2 cm left), suggestive of malignant infiltration. Adrenal protocol imaging revealed indeterminate lesions with attenuation >40 Hounsfield Unit and relative washout <40%. Ultrasound-guided biopsy of the right adrenal gland was performed.

The patient was discharged on hydrocortisone replacement with plans for early follow-up. One week later, he presented again in adrenal crisis with severe hypoglycemia (1.1 mmol/L) and cardiovascular collapse. Despite resuscitative efforts, he succumbed. Histopathology subsequently confirmed diffuse large B-cell lymphoma, activated B-cell subtype, consistent with PAL.

CONCLUSION
This case highlights the diagnostic pitfalls of PAL, particularly when constitutional symptoms mimic chronic infections such as tuberculosis. It underscores the need for high clinical suspicion in patients with bilateral adrenal masses and adrenal insufficiency. Early recognition, adequate steroid replacement including mineralocorticoid therapy, and prompt tissue diagnosis are critical to prevent rapid deterioration and improve outcomes.

EP_A015

METASTATIC MALIGNANT PHEOCHROMOCYTOMA DRIVEN BY DNMT3A SOMATIC MUTATION

Vijayrama Rao Sambamoorthy[1], Zanariah Hussein[1], Anthony Louis Kindu[2]
[1]Endocrine Unit, Medical Department, Hospital Putrajaya
[2]Nuclear Medicine Department, Institut Kanser Negara

INTRODUCTION
Pheochromocytomas and paragangliomas (PPGL) are rare neuroendocrine tumors with high heritability. While most are benign, approximately 25% are malignant, defined by distant metastases. Molecular classification has identified three clusters, with Cluster 3 (Wnt-signaling) being exclusively somatic and associated with aggressive behavior. We report a rare case of metastatic malignant pheochromocytoma driven by a somatic DNMT3A mutation, highlighting its unique imaging characteristics and rapid clinical progression.

CASE
A 55-year-old female presented with paroxysmal hypertension, headache, and a 10-kg weight loss. Biochemical workup revealed markedly elevated 24-hour urine metanephrines (163 × ULN) and normetanephrines (47 × ULN). Imaging confirmed a 15-cm left adrenal mass with liver and widespread skeletal metastases. Functional imaging demonstrated a striking mixed avidity: liver metastases were predominantly fluorodeoxyglucose-avid (SUVmax 7.0), skeletal lesions showed high Ga-68 DOTATATE avidity (SUVmax 6.9), and the primary tumor exhibited the strongest avidity on 131 I-MIBG scan. Whole Exome Sequencing identified a rare pathogenic somatic variant in the DNMT3A gene (c.2645G>A) with no other germline or somatic mutations in known susceptibility genes. Despite adequate alpha-blockade and supportive care, the patient developed acute liver failure and coagulopathy, rendering her unfit for any form of invasive intervention and finally succumbing to the disease within 3 months of presentation.

CONCLUSION
This case underscores the aggressive nature of Cluster 3 PPGLs associated with DNMT3A mutations, which likely promote tumorigenesis via Wnt-pathway activation and epigenetic dysregulation. The discordant functional imaging reflects significant tumor heterogeneity, which may complicate diagnostic and therapeutic strategies.