repeat imaging two months later demonstrated rapid progression, with diffusely scattered and enlarging liver metastases throughout both lobes (largest measuring 7.9 x 16.8 x 15.2 cm).

The patient experienced frequent, severe hypoglycemic episodes requiring prolonged hospitalization, high-concentration dextrose administration via multiple central venous catheters, high-dose corticosteroids, and further escalation of medical therapy. Over the course of his hospitalization, he developed recurrent sepsis and multi-organ dysfunction, ultimately leading to his death.

CONCLUSION

This case illustrates the aggressive nature, management complexities and therapeutic challenges of metastatic insulinomas. Several studies demonstrated that early administration of systemic chemotherapy in high-grade insulinomas has been associated with improved survival. Early consideration of advanced therapies like everolimus, PRRT and chemotherapy may be crucial in managing malignant insulinomas.

EP_A126

A RARE ENCOUNTER: HIRSUTISM UNMASKING ADRENAL ONCOCYTIC NEOPLASM IN A YOUNG WOMAN

https://doi.org/10.15605/jafes.040.S1.134

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INTRODUCTION/BACKGROUND

Adrenal oncocytic neoplasms (AONs) are rare tumors, with fewer than 300 cases reported since their first description in 1986. Most AONs are benign, non-secretory, and discovered incidentally. Hormone-secreting AONs are exceptionally uncommon. We present a case of a testosterone- and cortisol-secreting AON in an 18-year-old woman with primary amenorrhea and hirsutism.

CASE

An 18-year-old female presented with increased hair growth and primary amenorrhea. She had a history of unsuccessful hormonal therapy for amenorrhea since age 15. Physical examination revealed signs of hyperandrogenism, including hirsutism (Ferriman-Gallwey score 19), androgenic alopecia, deepened voice, and clitoromegaly. Pelvic ultrasound showed a small uterus with non-visualized ovaries. Laboratory investigations revealed elevated hematocrit (56%) and hormonal profiles indicative of hyperandrogenism and hypercortisolism. Abdominal computed tomography (CT) identified a 7.5 cm right adrenal mass with heterogeneous enhancement. A provisional diagnosis of a cortisol- and androgen-secreting adrenal tumor was made.

The patient underwent open right adrenalectomy with perioperative steroid coverage. Gross pathological examination was consistent with an AON. The tumor exhibited capsular and sinusoidal invasion but lacked vascular invasion, aberrant mitosis or necrosis. Based on Lin-Weiss-Bisceglia criteria, the tumor was classified as an AON of uncertain malignant potential.

Postoperatively, the patient experienced spontaneous menstruation five months after surgery. Follow-up CT at 15 months showed no recurrence or metastases, and hormonal profiles showed resolution of hyperandrogenism and hypercortisolism.

CONCLUSION

This case highlights a rare functional AON presenting with hyperandrogenism and hypercortisolism. Experienced pathologists play a crucial role in aiding accurate diagnosis. Complete surgical excision led to hormonal resolution and menstrual recovery, reinforcing the importance of considering adrenal tumors in young women with unexplained hyperandrogenism and amenorrhea.

EP_A127

AORTOCAVAL PARAGANGLIOMA IN VON HIPPEL-LINDAU DISEASE: A RARE EXTRA-ADRENAL PRESENTATION WITH DISTINCT BIOCHEMICAL AND CLINICAL PROFILE

https://doi.org/10.15605/jafes.040.S1.135

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INTRODUCTION/BACKGROUND

Pheochromocytomas and paragangliomas (PPGLs) are catecholamine-secreting tumors derived from chromaffin cells, with approximately 40% linked to germline mutations. One of the most common genetic associations is Von Hippel-Lindau (VHL) disease. VHL-related PPGLs typically arise in

the adrenal glands, with only 10–20% occurring in extra-adrenal sites. Here we describe a patient who presented with VHL-associated aortocaval paraganglioma.

CASE

A 34-year-old female with poorly controlled hypertension on triple therapy presented with paroxysmal symptoms and worsening renal function since December 2023. She was admitted in May 2024 for hypertensive urgency and renal impairment. Ultrasonography and CT scan of the abdomen revealed a 6 cm retroperitoneal aortocaval mass. Biochemical tests confirmed elevated normetanephrine levels with normal metanephrine levels, and Ga-68 DOTATATE PET/CT imaging showed a somatostatin receptor-avid paraganglioma. Her family history was notable for a sibling with pheochromocytoma. She received adequate alpha-blockade, followed by successful surgical excision of the tumour. Histology confirmed a paraganglioma with low proliferative activity (Ki-67 <1%). Genetic testing revealed a VHL missense variant, confirming a diagnosis of VHL disease. Subsequent surveillance for other VHL-related manifestations revealed no additional tumours, and she is currently in remission for PPGL. Family screening identified five other individuals, including her young son, with the same genetic mutation; all are now undergoing regular follow-up.

CONCLUSION

VHL-associated PPGLs present at a younger age than sporadic cases and primarily secrete noradrenaline due to reduced PNMT expression, resulting from impaired hypoxic pathways caused by VHL loss. Patients often present with chronic hypertension and tachycardia. Functional imaging with 18F-DOPA PET/CT is preferred for its high sensitivity. These tumors have a low metastatic risk (5–8%) and rarely require systemic therapy. Given the high mutation penetrance of VHL disease (~90% by age 65), lifelong surveillance is essential. Early genetic and clinical monitoring enables timely detection in patients and at-risk relatives.

EP_A128

PERMANENT CENTRAL DIABETES INSIPIDUS IN A POST TRANSSPHENOIDAL SURGERY PATIENT: A CASE REPORT

https://doi.org/10.15605/jafes.040.S1.136

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INTRODUCTION/BACKGROUND

Diabetes insipidus (DI) is a disorder characterized by the excretion of large volumes of hypotonic urine. Four types of DI must be differentiated: central DI (cDI), nephrogenic DI (nDI), gestational DI, and primary polydipsia. Central DI can be transient, particularly as a complication of pituitary surgery. Permanent central DI is a rare complication of pituitary surgery.

CASE

A 48-year-old female was referred from the neurosurgery department with a nine-month history of polyuria (urination exceeding 3 liters daily), polydipsia (excessive thirst), fatigue, constipation, and cold intolerance. Nine months prior, she underwent transsphenoidal surgery for a pituitary macroadenoma. She reported no prior history of hormonal abnormalities or use of antithyroid medication. Her body mass index (BMI) was 24.5 kg/m^2, and her thyroid gland was not enlarged.

Laboratory results showed: TSH 0.25 mIU/mL, FT4 9.5 pmol/L, cortisol 312 µg/dL, prolactin 268 ng/mL, LH 2 mIU/mL, FSH 11.35 mIU/mL, estradiol 62.3 pg/mL, sodium 143 mmol/L, urine osmolality 100 mOsm/kg, serum osmolality 292 mOsm/kg. A water deprivation test (WDT) revealed a urine osmolality of 212 mOsm/kg, which increased to 499 mOsm/kg after desmopressin administration.

The diagnoses were central diabetes insipidus (cDI) (post-operative, permanent), central hypothyroidism, and hypogonadotropic hypogonadism. Treatment consisted of desmopressin 0.6 mg once daily and thyroxine 50 mcg once daily. After six months, her signs and symptoms improved.