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.

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

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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.

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

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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