Adult E-Poster

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METASTATIC PULMONARY NEUROENDOCRINE NEOPLASM WITH CARCINOID SYNDROME COMPLICATED BY BOWEL PERFORATION

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INTRODUCTION/BACKGROUND
Carcinoid syndrome (CS) results from hormone-secreting neuroendocrine neoplasms (NENs) releasing bioactive substances into systemic circulation. NENs are most commonly found in the gastrointestinal tract and less frequently in the lungs. CS develops in about 19% of patients with NENs, with 20% presenting with distant metastases. Rarely, mesenteric fibrosis in CS can lead to ischemia and perforation.

We present a case of metastatic bronchial NEN with CS, complicated with bowel perforation and subsequent death.

CASE
A 46-year-old male with previous history of spinal surgery for a post-traumatic compression fracture presented with gradual bilateral lower limb weakness and back pain, followed by chronic diarrhea and significant weight loss. Spinal MRI revealed extensive metastatic bone disease. Oesophagogastroduodenoscopy (OGDS) and colonoscopy were unremarkable; however, CT imaging identified a solitary endobronchial mass in the left lower lobe (3.1 x 2.6 x 2.8 cm), associated with left hilar lymphadenopathy and liver metastases.

Biopsy of the lung mass revealed a grade 1 neuroendocrine tumor (Ki67 1%). Urinary 5-hydroxyindolacetic acid (5-HIAA) levels were markedly elevated at 854.6 µmol/day. Gallium-68 DOTATATE PET-CT demonstrated somatostatin receptor-avid disease involving the left lung, with mediastinal nodes, liver and extensive skeletal metastasis. A diagnosis of CS was established based on clinical presentation, elevated 5-HIAA, imaging, and histopathology.

The patient was initiated on octreotide, a somatostatin analogue. However, he struggled to come to terms with the diagnosis and self-discharged against medical advice. He was later readmitted with severe hypokalemia, acute kidney injury, metabolic acidosis and acute abdomen. CT imaging revealed pneumoperitoneum consistent with a perforated duodenum. Due to hemodynamic instability, surgical intervention was not feasible, and palliative care was given.

CONCLUSION
This case illustrates a rare and potentially fatal complication of CS, underscoring the importance of early diagnosis and prompt treatment. Maintaining a high index of suspicion is crucial for timely identification of CS.

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TRAPPED IN THE HYPOGLYCEMIA LOOP: A RARE CASE OF RAPIDLY PROGRESSIVE METASTATIC INSULINOMA

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INTRODUCTION/BACKGROUND
Malignant insulinomas are rare and account for approximately 10-15% of all insulinomas. Most metastatic insulinomas are not curable with surgery alone and necessitate a multimodal approach encompassing medical, locoregional, targeted, systemic, and supportive therapies. The optimal treatment sequence should be individualized to each patient.

CASE
A 21-year-old male presented with a one-month history of recurrent hypoglycemic episodes characterized by neuroglycopenic symptoms. Subsequent evaluation confirmed endogenous hyperinsulinemic hypoglycemia. Computed tomography (CT) imaging revealed a 2.1 x 2.0 x 1.8 cm lesion in the pancreatic tail, multiple liver lesions in both lobes (largest measuring 3.8 x 4.1 x 4.1 cm), and intra-abdominal lymphadenopathy (largest measuring 1.1 x 1.8 cm). Gallium-68 (Ga-68) DOTATATE PET/CT and fluorine-18 fluorodeoxyglucose (18F-FDG) PET/CT demonstrated predominantly somatostatin receptor-avid disease.

Following multidisciplinary team discussion, the tumor was deemed inoperable. Medical management was rapidly escalated, involving diazoxide, hydrochlorothiazide, corticosteroids, octreotide followed by pasireotide, and dextrose infusion, guided by continuous glucose monitoring. Endoscopic ultrasound-guided fine needle biopsy (EUS-FNB) of the pancreatic tail lesion revealed a high-grade, well-differentiated neuroendocrine tumor (Ki67 50%, G3).

While awaiting access to systemic therapies, including everolimus, peptide receptor radionuclide therapy (PRRT), and chemotherapy, the patient underwent radiofrequency ablation (RFA) of the pancreatic tail lesion and transarterial embolization (TAE) of the hepatic lesions. However,

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

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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

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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