

Adult E-Poster

A second course of IV liposomal Amphotericin B was administered, followed by itraconazole. The patient's fever resolved, and follow-up imaging showed reduction in adrenal mass size. He remains on drainage and long-term antifungal therapy, planned for at least 18 months.

CONCLUSION

This case highlights the diagnostic challenges of adrenal histoplasmosis in immunocompetent individuals presenting with vague systemic symptoms and large bilateral adrenal masses. Early recognition and a multidisciplinary approach are crucial for timely diagnosis and optimal management.

EP_A162

BEYOND THE YELLOW: UNMASKING PHEOCHROMOCYTOMA IN A JAUNDICED PATIENT

<https://doi.org/10.15605/jafes.040.S1.170>

Seetha Devi Subramanian,¹ Gerard Jason Mathews,¹ Nor Shaffinaz Yusoff Azmi Merican,¹ Nadiyah Ahmad Sabri,² Shartiyah Ismail¹

¹Endocrinology Unit, Department of Medicine, Hospital Sultanah Bahiyah, Kedah, Malaysia

²Anatomy Pathologic Unit, Department of Pathology, Hospital Sultanah Bahiyah, Kedah, Malaysia

INTRODUCTION/BACKGROUND

Pheochromocytomas and paragangliomas (PPGLs) are rare neuroendocrine tumours (NET) arising from chromaffin cells. Bilateral pheochromocytomas are extremely rare, constituting 7–10% of all pheochromocytoma cases, and 60%–90% of them possess a germline mutation.

CASE

A 17-year-old male presented initially with epigastric pain and obstructive jaundice. He is not hypertensive. ERCP revealed choledocholithiasis and a dilated common bile duct (CBD). A contrast-enhanced CT of the liver showed an enhancing CBD lesion causing biliary obstruction and incidental bilateral adrenal tumours. A CT Adrenal protocol confirmed a left adrenal lesion measuring 5.4×4.8×5.1cm with unenhanced attenuation 35.4 Hounsfield Units (HU) and a right adrenal lesion measuring 1.2 × 1.1 × 1.6 cm with unenhanced attenuation 30.7HU, both with delayed contrast washout, consistent with pheochromocytomas. Biochemical evaluation showed elevated 24-hour urinary normetanephrine at 18,558 nmol/24h (497–2489), which is seven times the upper limit of normal, confirming catecholamine excess with normal levels of Metanephrine. Other hormonal investigations were unremarkable.

He underwent open cholecystectomy and choledochectomy with biliary reconstruction. Histopathology confirmed a well-differentiated Grade I neuroendocrine tumour (NET) of the CBD, positive for synaptophysin, chromogranin A, and CD56, with a Ki-67 <3%. Surgical margins and lymph nodes were negative. Thyroid ultrasound was normal. ⁶⁸Ga-DOTATATE Positron Emission Tomography (PET) confirmed bilateral pheochromocytomas with no extra-adrenal paraganglioma or metastatic disease. He underwent bilateral adrenalectomy after adequate alpha-blockade and was discharged well with hydrocortisone and fludrocortisone replacement. He is awaiting genetic testing.

CONCLUSION

This case highlights a rare pheochromocytoma with obstructive jaundice, lacking the classical triad of headache, palpitations, and sweating. Bilateral pheochromocytomas are commonly seen in Multiple Endocrine Neoplasia types 2A and 2B, von Hippel–Lindau disease, and rarely with MAX and TMEM127 mutations, though they can also occur sporadically. Genetic testing is crucial for diagnosing and managing bilateral pheochromocytoma, as it aids in treatment decisions, recurrence prediction, and family screening.

EP_A163

TREACHEROUS JOURNEY OF ADVANCED PAPILLARY THYROID CARCINOMA IN PREGNANCY

<https://doi.org/10.15605/jafes.040.S1.171>

Seetha Devi Subramanian, Gerard Jason Mathews, Tan Jie En, Noor Rafhati Adyani Abdullah, Shartiyah Ismail, Nor Shaffinaz Yusoff Azmi Merican

Endocrinology Unit, Department of Medicine, Hospital Sultanah Bahiyah, Kedah, Malaysia

INTRODUCTION

Papillary thyroid carcinoma (PTC) is the most common thyroid malignancy and generally exhibits a favourable prognosis, but it can manifest with metastasis in advanced stages. Pregnancy complicates the management of such cases particularly when radioactive iodine (I-131) therapy is indicated.

CASE

A 26-year-old presented in her second trimester with acute exacerbation of bronchial asthma requiring mechanical ventilation. During intubation, a 6 × 4 cm anterior neck swelling was found. A computed tomography showed diffuse heterogeneous thyroid enlargement with tracheal narrowing, cervical lymphadenopathy, and pulmonary

Adult E-Poster

metastasis. Thyroid function tests were normal. She underwent debulking thyroidectomy with bilateral modified radical neck dissection. Patient required a tracheostomy due to tumour invasion into the trachea. Histopathology confirmed multifocal (>5 foci) classical variant PTC with the largest nodule measuring 25 mm. The tumour showed lymphovascular invasion, regional nodal metastases, and invasion into adjacent skeletal muscle indicating an advanced stage with a high risk of recurrence as per the American Thyroid Association (ATA) risk stratification.

Postoperatively, we started her on levothyroxine with a TSH target of below 0.1mIU/L. At 33 weeks gestation, an elective lower segment caesarean section was performed. Post delivery, cabergoline was given to suppress lactation in preparation for I-131 therapy. After consultation with nuclear medicine, I-131 therapy was scheduled at 10 weeks postpartum. Levothyroxine was withheld one month prior.

CONCLUSION

This case highlights the challenges of managing advanced PTC with metastasis during pregnancy. Thorough multidisciplinary planning of surgery and postpartum I-131 timing is essential to ensure a seamless delivery and safety of mother and child. To safeguard breast tissue from radiation exposure, breastfeeding should be entirely discontinued at least six weeks prior to I-131 therapy. Breastfeeding should not be resumed after I-131 administration to shield the infant from radiation exposure and avert harm to the infant's thyroid gland. Breastfeeding is not contraindicated in subsequent pregnancies.

EP_A164

TEMOZOLOMIDE THERAPY IN RECURRENT METASTATIC PHEOCHROMOCYTOMA: A CASE-BASED REVIEW

<https://doi.org/10.15605/jafes.040.S1.172>

Hidayatil Alimi Bin Keya Nordin, Tong Chin Voon, Zanariah Binti Hussein

Institut Endokrin, Hospital Putrajaya, Putrajaya, Malaysia

INTRODUCTION

Metastatic pheochromocytoma is rare and the management is complex, requiring multifaceted, multidisciplinary management. Primarily palliative, treatment focuses on tumor control, symptom management, and quality of life. While historically associated with a poor prognosis, improved diagnosis and management, including surgery, chemotherapy and targeted therapies, are extending survival for some patients.

CASE

A 52-year-old female was initially diagnosed in 2013 with non-functioning pheochromocytoma with liver, spleen, and pancreatic tail metastases. She underwent left adrenalectomy, splenectomy, distal pancreatectomy, and local resection of metastatic liver lesions, followed by trans-arterial chemoembolization of liver metastases. Subsequent follow-up imaging revealed recurrent disease, necessitating further surgical intervention. This included left hemihepatectomy, left nephrectomy, segmental resection of the colon and splenic flexure and excision of a posterior abdominal tumor. Due to the extensive nature of her disease progression, the patient received 4 cycles of peptide receptor radionuclide therapy (PRRT) as well as palliative radiotherapy to left thoracoabdominal mass and T9 till L1 vertebrae. Despite undergoing PRRT, the disease continued to progress. A multidisciplinary team discussion led to the initiation of temozolomide treatment in March 2023. The patient has received 22 cycles of temozolomide from 2023 to date, with recent follow-up imaging demonstrating partial response to the treatment.

CONCLUSION

This case report illustrates therapeutic efficacy of temozolomide in metastatic pheochromocytoma. In recent years, temozolomide has shown good outcomes in some metastatic pheochromocytoma patients, especially those with SDHB germline mutation. Temozolomide treatment has been generally considered to have a low toxicity profile, however few studies have noted the development of severe myelosuppression. While the current evidence base is still developing and primarily relies on retrospective data and case reports, ongoing clinical trials are anticipated to yield more definitive conclusions regarding its efficacy and optimal clinical application in metastatic pheochromocytoma.