

A 73-year-old Chinese male with a history of type 2 diabetes mellitus, hypertension and stage IV chronic kidney disease presented to emergency department with syncope attack and capillary blood glucose of 1.9 mmol/L. Initial examination revealed massive hepatomegaly. A 4-phase CT scan of the liver showed multicentric hepatocellular carcinoma. Biochemical investigations revealed hypoinsulinaemic hypoglycaemia, elevated alpha-fetoprotein (AFP) at 135,937 IU/mL, markedly suppressed insulin-like growth factor-1 (IGF-1) and normal IGF-2 level. The IGF-2: IGF-1 ratio was 50:1. He was started on oral prednisolone, titrated up to 25mg twice daily to maintain euglycemia. After discussion with surgical and oncology teams, patient opted for conservative management.

Elevated IGF-2 or pro-IGF-2 exerts insulin mimicking effects leading to hypoglycaemia. The diagnosis of NICTH is based on the IGF-2: IGF-1 ratio, which is higher than 10:1, along with inappropriately suppressed IGF-1. Curative treatment for NICTH is complete resection of the tumour. Glucocorticoids is important to maintain euglycemia on top of nutritional support and glucose infusion. Glucagon has a limited role as adjunct therapy. Diazoxide and octreotide were found ineffective.

CONCLUSION

This is a case of hepatoma-associated NICTH, which was managed with oral prednisolone to maintain euglycemia. Due to advanced disease, we were unable to deliver definitive treatment. High level of suspicion of NICTH is crucial in patients with recurrent hypoglycaemia on a background of malignancy.

EP_A036

RENINOMA: A SURGICALLY CURABLE CAUSE OF HYPERTENSION

<https://doi.org/10.15605/jafes.038.S2.54>

Gaayathri Krishnan

Endocrinology, Ministry of Health, Malaysia (KKM)

INTRODUCTION/BACKGROUND

Reninoma is an extremely rare cause of hypertension. It is a tumour of the juxtaglomerular apparatus which secretes renin, leading to secondary hyperaldosteronism, ultimately causing hypertension. This disease is commonly seen in adolescents and young adults and has a female preponderance. Patients with reninoma mostly present with hypokalaemia although they can also be normokalaemic. Most reninomas are benign and surgical resection can render the patient normotensive. Here we report a case of reninoma in a young female who presented with hypertension and palpitations.

CASE

We describe a 23-year-old female who was referred for evaluation of hypertension in the young. She was found to have a BP ranging between 130-180 mmHg (systolic) and 85-120 mmHg (diastolic). She was diagnosed with hypertension and started on oral doxazosin. She complained of paroxysms of sweating with palpitations. On examination, she was a moderately built female with a body mass index of 20.4kg/m². Her pulse rate was 100 beats per minute and her BP was 153/100 mmHg.

Her renin levels were elevated at 518 mU/L (NV: 4.4- 46.1 mU/L) with elevated aldosterone at 998 pmol/L (NV: 61.2- 997.8 pmol/L). Computed tomography of abdomen and pelvis revealed the presence of a well encapsulated heterogeneously enhancing mass on the upper pole of the right kidney measuring 2.7 x 3.0 x 3.3 cm. A biopsy of the lesion was suggestive of a reninoma. The patient underwent a nephron-sparing surgery. Histopathological examination revealed an encapsulated lesion that stained positive for CD 34, CD 177 and vimentin, confirming the diagnosis. Following surgery, the patient was normotensive and all anti-hypertensive medications were withheld.

CONCLUSION

Reninomas are mostly benign neoplasms that can lead to hypertension and severe end-organ damage. High clinical suspicion is required to diagnose this disease, and nephron-sparing surgery can render the patient normotensive.

EP_A037

A BONY PREDICAMENT: CATCHING THE CULPRIT IN THE CHEST

<https://doi.org/10.15605/jafes.038.S2.55>

Quan Hziung Lim, Nicholas Ken Yong Hee, Ken Seng Chiew, Marisa Khatijah Borhan, Katherine Khor, Ee Wen Loh, Tharsini Sarvanandan, Sharmila Paramasivam, Lee Ling Lim, Jeyakantha Ratnasingam, Siew Pheng Chan, Shireene Ratna Vethakkan

Endocrinology Unit, Department of Medicine, Universiti Malaya Medical Centre, Kuala Lumpur, Malaysia

INTRODUCTION/BACKGROUND

We report a case of mediastinal parathyroid adenoma as a rare cause of severe hypercalcemia and bone deformities.

CASE

A 19-year-old male presented with progressively worsening scoliosis and restrictive chest wall deformity over 4 years. He underwent chest wall reconstruction surgery with insertion of a titanium plate. Hypercalcemia was incidentally

diagnosed perioperatively. He had polydipsia and weight loss but denied other symptoms. Past medical history and family history were unremarkable. He was small in stature with a body mass index (BMI) of 12.6 kg/m², severely scoliotic spine and pectus excavatum and no palpable neck masses.

Skeletal survey showed multiple malunited fractures and lytic bone lesions. Biochemical investigations revealed severe hypercalcemia with a peak level of 4.38 mmol/L and a markedly elevated intact PTH (iPTH) level of 695.3 pmol/L. Initial technetium-99m-sestamibi (MIBI) parathyroid scintigraphy showed no uptake over the neck and neck ultrasound revealed no masses. A subsequent whole-body MIBI with SPECT-CT showed avid uptake at the anterosuperior mediastinum, corresponding to a contrast-enhancing mass seen on CT-scan, measuring 17 x 30 x 18 mm. Genetic screening was negative for mutations in CASR, MEN1, MEN2 and MEN4 genes.

Hypercalcemia was controlled with subcutaneous denosumab and oral cinacalcet before subsequent surgical resection. The mediastinal mass removed measured 55 mm in diameter and weighed 12.5 grams. Histopathological examination revealed tissue predominantly comprising chief cells, confirming an ectopic PTH adenoma. Post-operatively, his iPTH levels normalized and he required high doses of alfacalcidol.

CONCLUSION

Mediastinal adenomas are ectopic parathyroid adenomas that form because of aberrant embryological migration of inferior parathyroid glands. They may pose a diagnostic challenge leading to unnecessary surgical explorations. MIBI scans can improve diagnostic accuracy. Ectopic parathyroid adenomas make up 16-22% of parathyroid adenomas, of which 6-13% are mediastinal adenomas. Patients with primary hyperparathyroidism and initial negative neck imaging should prompt suspicion and work-up for mediastinal parathyroid adenoma.

EP_A038

PRIMARY HYPERPARATHYROIDISM IN PREGNANCY, INITIAL PRESENTATION WITH HYPOCALCAEMIA SECONDARY TO ACUTE PANCREATITIS AND SEPSIS

<https://doi.org/10.15605/jafes.038.S2.56>

Alvin Gnanasegaran and Sivasangkari Mugilarasan

Endocrine Unit, Department of Medicine Hospital Taiping, Malaysia

INTRODUCTION/BACKGROUND

Diagnosis of primary hyperparathyroidism is challenging during pregnancy and requires special care to prevent complications such as nephrolithiasis, pancreatitis, preterm delivery and foetal demise. We are reporting a rare case of primary hyperparathyroidism initially presenting with severe hypocalcaemia.

CASE

A 25-year-old primigravida at 24 weeks of gestation was admitted to intensive care unit with decompensated septic shock and acute kidney injury requiring ventilatory support. Subsequently, she had chorioamnionitis and non-viable foetus resulting in premature delivery. CECT of the abdomen showed acute pancreatitis in the absence of gallstone and caecal perforation with extensive peritonitis. She then underwent laparotomy. Other laboratory results showed elevated white blood count (29.7 g/d) and serum creatinine (173 micromol/L) with reduced eGFR (35 ml/mol/1.73 m²), hypocalcaemia (1.79 mmol/L (corrected)), hypoalbuminemia (21 g/L), with normal phosphate (0.91 mmol/L). She required multiple intravenous calcium corrections for the first 3 days. The serum calcium showed gradual increment from 2.4 mmol/l to 4.07 mmol/L within 10 days without calcium or vitamin D supplementation and adequate hydration. Intact parathyroid hormone (iPTH) was 43 pmol/L but repeat test was elevated at 148 pmol/L. She required intravenous zoledronate 4 mg with bridging calcitonin given 3 weeks apart. She had severe vitamin D deficiency of <5 ng/mL for which she was started on replacement. Patient denied familial hypercalcaemia or MEN syndrome. Ultrasound of the neck did not locate a parathyroid adenoma. Therefore, she is awaiting sestamibi scan and genetic testing to rule out familial causes of primary hyperparathyroidism is being considered.

CONCLUSION

Hypocalcaemia is a common finding in acute pancreatitis due to mesenteric calcium salt formation. It is also present in critically ill patients with sepsis which is a marker of severity with increased mortality and hospital stay.