impairment in the left ear, an investigation for suspicion of glomus tympanicum commenced 6 months later. He had repeated ER visits for the next year due to the persistence of symptoms and uncontrolled hypertension. However, he was only referred to the endocrine clinic for young hypertension investigation 20 months from the initial presentation with significant paroxysms of palpitation, headaches and elevated BP. Diagnosis of catecholamine-secreting glomus-jugulotympanicum-paraganglioma was confirmed with elevated urinary metanephrine and huge soft tissue mass in the left jugular fossa with local bony erosion and intracranial extension on MRI.

His BP control was labile and required multiple oral-antihypertensives including phenoxybenzamine. Multi-disciplinary team management prepared him for definitive surgical intervention. He underwent tumour embolization prior to the actual surgery. Pre-operative management was extremely challenging which required CCU admission for BP stabilization. Intra-operative period was surprisingly uneventful, but he developed multiple cranial nerve palsies postoperatively. A second operation was required due to infection and enlarging tumor with compression. Paroxysm of symptoms improved after second surgery but he still had significant residual tumor. MIBG-therapy was planned but management was delayed due to the COVID-19 pandemic and treatment funding.

CONCLUSION

Awareness of functional paraganglioma presentation is imperative to avoid late detection. HNPGLs that are both aggressive and functional pose extreme difficulty in achieving disease remission.

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ADIPSIC CENTRAL DIABETES INSIPIDUS IN A PATIENT WITH SUPRASELLAR GERMINOMA

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INTRODUCTION/BACKGROUND

Adipsic central diabetes insipidus (CDI) is a rare and life-threatening disease which is difficult to manage as the patient experiences loss of thirst sensation compounded by fluid loss and hypernatremia. We present a case of a patient with suprasellar germinoma with panhypopituitarism who presented with adipsic CDI.

CASE

A 20-year-old male presented with generalized body lethargy for 1 month and polyuria for 1 year. He did not complain of polydipsia. Blood tests revealed severe hypernatremia with serum sodium (Na) of 160 mmol/L, serum osmolality of 300 mOsm/kg and urine osmolality of 130 mOsm/kg. Other electrolytes and blood glucose were normal. MRI showed a large sellar/suprasellar mass with periventricular subependymal spread causing acute obstructive hydrocephalus. Hormonal panel showed panhypopituitarism. He was started on sublingual desmopressin, L-thyroxine, hydrocortisone and intramuscular testosterone. During his confinement, he denied polydipsia despite intermittent polyuria. Strict intake and output monitoring were instituted with hourly urine output and regular renal profile monitoring. Despite initial normalisation of Na levels, he developed 2 episodes of hypernatremia when he had breakthrough polyuria. Intravenous fluids were given intermittently to balance his output. The patient and caregiver were constantly reminded to take adequate oral fluid. He then underwent transcranial biopsy. Histopathology examination showed a diagnosis of central nervous system germinoma. A referral to the Oncology team for chemotherapy was made.

Adipsic CDI has been reported to account for about 10% of CDI cases. Patients with adipsic CDI have higher prevalence of complications such as hypernatremia, renal insufficiency and venous thrombosis. Apart from desmopressin, crucial management steps include regular monitoring to ensure adequate fluids and desmopressin replacement.

CONCLUSION

This case highlights the difficulties in managing adipsic CDI and the need for constant and regular monitoring to prevent life-threatening hypernatremia.