P88

Multiple Endocrine Neoplasia Type 1 Presenting as Insulinoma, Parathyroid Adenoma and Multiple Collagenomas

Dr Edelissa Payumo[1], Dr Jonathan Mercado[1], Dr James Paningbatan[1], Dr Ma. Leonora Capellan[1]
[1]*Makati Medical Center , Makati, Philippines*

Background: Multiple Endocrine Neoplasia Type 1 (MEN1), a rare autosomal dominant disorder, can be diagnosed clinically by the presence of at least two primary MEN1-associated tumors that include parathyroid adenoma, enteropancreatic tumor, and pituitary adenoma. Cutaneous tumors are also common in this syndrome. We report a case of MEN1 presenting with insulinoma, parathyroid adenoma, and multiple collagenomas.

Case: A 43-year-old male had history of recurrent episodes of generalized body weakness associated with dizziness, diaphoresis, and tremors for six years, relieved with food intake. He had no family history of MEN1. He was obese and with notable multiple skin-colored papulonodular skin lesions on the abdomen, back and extremities. He had a low fasting serum glucose of 41.72 mg/dl with unsuppressed serum insulin and C-peptide consistent with endogenous hyperinsulinemia. Contrast MRI of the abdomen revealed a 2 x 1.8 cm nodule within the pancreatic tail. Biopsy of skin lesion was consistent with collagenoma. The occurrence of both insulinoma and collagenoma in this case prompted work up for MEN1 which also revealed primary hyperparathyroidism. This was confirmed by a Sestamibi scan consistent with parathyroid adenoma. Screening for pituitary involvement was negative. Patient subsequently underwent staged distal pancreatectomy with splenectomy and subtotal parathyroidectomy. There was resolution of hyperinsulinemia and hyperparathyroidism postoperatively. Histopathology report was neuroendocrine tumor of the pancreas and parathyroid hyperplasia.

Conclusion: In the absence of a family history and genetic testing, screening with biochemical parameters and imaging is valuable in the diagnosis of MEN1 in case of a high index of suspicion.