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Internal Medicine Section

An Unusual Presentation of a Patient with Multiple Endocrine Neoplasia- 1

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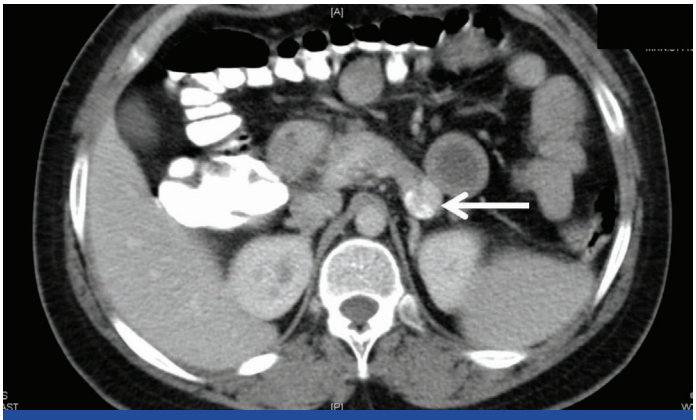
INTRODUCTION

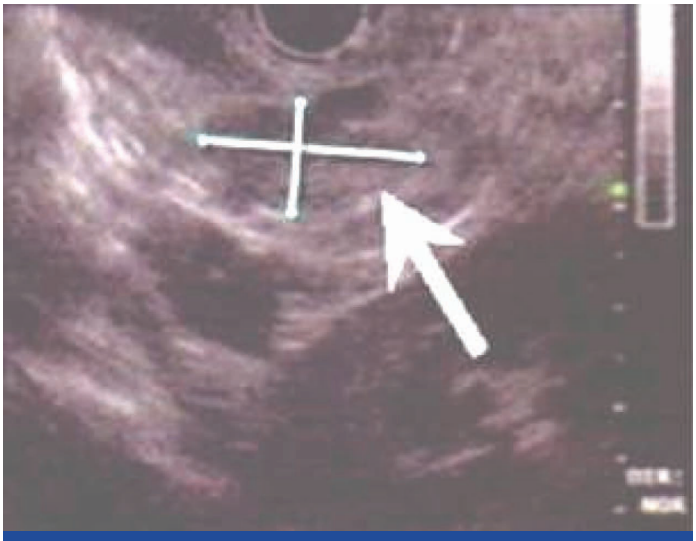
Prolonged under-recognized hypoglycaemia may later manifest with wide range of neurological symptoms and signs including recurrent seizures, cognitive decline, ataxia secondary to cerebellar atrophy, small muscle wasting and peripheral neuropathy. We hereby report a young man who presented with recurrent episodes of seizures and intention tremor.

CASE PRESENTATION

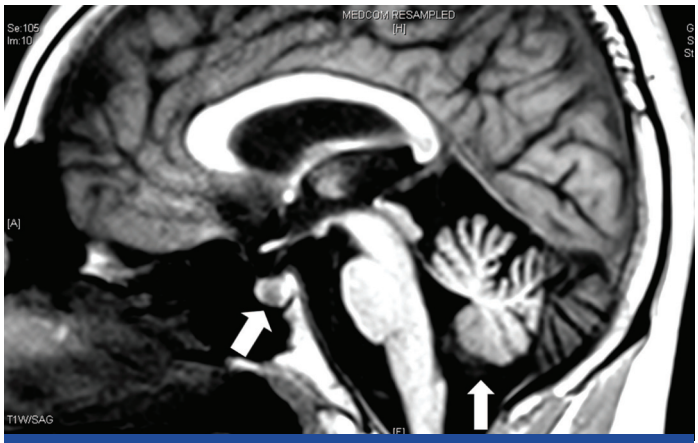
A 24-year old gentleman presented with multiple episodes of loss of consciousness, generalised seizures and intention tremors of his extremities of five years duration. He did not have headache or visual disturbances. There were no addictions. He was not on any regular medications. He was found to have a BMI of 28 kg/m^2 with bilateral cerebellar signs. Rest of the physical examination was unremarkable. On routine biochemical evaluation, he had random plasma glucose of 16 mg/dl in casualty. After stabilization, endocrinologist team decided to further scheduled for a 72 h fast, not only to confirm this unusually low value but also to document concomitant endogenous hyperinsulinemic hypoglycaemia. His random blood glucose prior to fast was 71 mg/dl. He developed hypoglycaemia within two hours of fast (glucose of 28 mg/dl) with a corresponding non suppressed serum insulin level of 15.2 units/ml characteristic of hyperinsulinemic hypoglycaemia. Further biochemical evaluation revealed an elevated serum prolactin of 161 ng/ml (Normal 2.5-17 ng/ml) indicating that he is having a prolactinoma, hypercalcaemia with albumin corrected calcium of 10.8 mg/dl (Normal 8.3-10.4 mg/dl) and also an elevated serum parathormone level of 185 pg/ml (Normal 8 -50 pg/ml) suggestive of primary hyperparathyroidism. A CT scanning of the abdomen revealed a well-defined ovoid exophytic lesion arising from the inferior aspect of the tail of pancreas measuring 23 X 17mm in the arterial phase [Table/Fig-1] which was also localized on an endoscopic ultrasound [Table/Fig-2]. He also underwent an MRI of the brain which displayed a pituitary microadenoma (prolactinoma) and a gross cerebellar atrophy [Table/Fig-3] secondary to prolonged unrecognised hypoglycaemia. His Sestamibi parathyroid scintigraphy was negative for an adenoma probably due to all 4 glands hyperplasia. Thus, this patient was diagnosed to have Multiple Endocrine Neoplasia Type1 (MEN1) with an insulinoma, primary hyperparathyroidism and a prolactinoma. The cerebellar atrophy was due to repeated untreated hypoglycaemic episodes. Multiple endocrine neoplasia type 1 (MEN-1) causes combinations of over 20 different endocrine and non-endocrine tumours, many of which could be benign or malignant and may involve the parathyroid, enteropancreatic neuroendocrine system or the anterior pituitary [1]. Since many of these tumours can be multicentric in origin a thorough screening is needed preoperatively to characterize the extent of disease and thereby prevent residual lesions and incomplete surgeries [2]. The MEN-1 syndrome is caused by mutations in the MEN-1 tumour suppressor gene located on



[Table/Fig-1]: CT Abdomen showing a lesion in the tail of the pancreas



[Table/Fig-2]: Endoscopic Ultrasound diplaying the mass



[Table/Fig-3]: MRI Brain showing a pituitary microadenoma and gross cerebellar atrophy

chromosome 11q13. The screening and identification of underlying genetic mutations will help in arriving at an early diagnosis and timely treatment of other components of the syndrome (MEN 1) in the index case and as well as in the family members. Management of these patients require multidisciplinary team approach including endocrinologists, surgeons and nuclear physicians. In non-diabetic subjects presenting with seizures, hypoglycaemia must be always be considered, as an early diagnosis and therapeutic measures will prevent the irreversible neuronal damage as seen in this patient.

Once a diagnosis of insulinoma is established, they should be further screened for other components of MEN1 and be managed by multidisciplinary team at an advanced tertiary care center.

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