ABSTRACT:

An insulinoma is a rare endocrine tumor of pancreas that causes hypoglycemia through its inappropriate secretion of insulin. It is a very rare tumor with annual incidence of 1-32 per million population (1). Insulinomas are usually benign tumors, sporadic and with a solitary small mass (<2cm diameter). (2) Insulinomas are found with equal frequency in head, body and tail of pancreas. Symptoms of hypoglycemia include both adrenergic symptoms (pallor, sweating, tremors and tachycardia) and neuroglycopenic symptoms (irritability, confusion, aggression, seizures and coma). Whipple’s triad should be confirmed as part of diagnostic workup, but be aware that over time, hypoglycemia unawareness may develop and potentially confound the clinical picture. Early localization of the disease is essential to prevent fatal hypoglycemia. The gold standard test for biochemical diagnosis includes plasma glucose, insulin, C-peptide and proinsulin during a 72 hours fasting. More than one imaging modality is usually required to localize the tumor, including computed tomography (CT), magnetic resonance imaging (MRI), endoscopic ultrasonography (EUS), intra-arterial calcium stimulation with hepatic venous sampling or angiography, and arterial stimulation venous sampling (ARVS). Diagnosis of this rare disease requires high index of suspicion based on clinical and laboratory findings and imaging to localize the lesion. Surgery is often curative and is the standard of care to prevent neurological deficit associated with prolonged hypoglycemia (3) though medical therapy can be used in the initial treatment.

CASE PRESENTATION:

24 years old male, medical student by profession, presented in endocrinology clinic with 6 months history of recurrent episodes if documented hypoglycemia, associated with hypoglycemic symptoms (both adrenergic and neuroglycopenic including one episode of fits), and improvement of symptoms after taking high glycemic indexed foods. He gained almost 20kg weight in last 6 months. There was no history of diabetes in the family or himself taking any antidiabetic medications. On examination, he had normal physical examination with slightly raised BMI of 25. Blood workup showed inappropriately high insulin level (13.6 u IU/ml) and high C-peptide levels (1.74ng/ml) with low blood sugar level of 25mg/dl and good cortisol response (28.97ug/dl, normal range: 4.30-22.4). Imaging studies including CT abdomen with contrast, MRI abdomen with contrast and DOTA PET scan did not reveal any structural lesion. EUS was planned to localize the lesion as it has high sensitivity as compared to CT or MRI. EUS showed a well-defined, ovoid mass measuring 19x8mm in the head of pancreas. He underwent Whipple’s procedure. Histopathology showed well-differentiated neuroendocrine tumor, WHO grade I, 1.7cm in size. Initially in the post-operative period he required small doses of insulin to control hyperglycemia, now he is off insulin and doing well.

PRACTICAL IMPLICATIONS:

Endoscopic ultrasound (EUS) should be considered as imaging modality of choice in cases of difficult to localize biochemically proven insulinomas. With early localization of structural lesion by using highly sensitive imaging modalities like EUS, we can prevent severe life threatening episodes of hypoglycemia by timely surgical removal.