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Case Report

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Seizure, Not Always A Referral for The Neurologist. A Case of Insulinoma Presenting as Confusion, Odd Behavior and Grand -Mal Seizure

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Abstract

Insulinoma is a rare neuroendocrine tumor of the pancreas that produces insulin in an uncontrolled fashion. Due to high circulating insulin levels in the blood, patients will often, if not most times, present with neurological symptoms. Symptoms can range in severity depending on the degree of hypoglycemia. In the most severe form, patients can experience seizures and coma. In less severe cases, they can present with fatigue, confusion, weakness, change in personality. Symptoms that resolve with food intake is a traditional hallmark. This hallmark diagnostic criteria should alert the health care provider to this diagnosis when a patient presents as such.

Introduction

Insulinoma is a very rare neuroendocrine tumor of the pancreas. It occurs in 0.5-5 per million people a year [1]. It is typically associated with MEN-1 syndrome, but it can occur sporadically as well. The tumor secretes insulin in an unregulated fashion. The normal bio feedback mechanism is interrupted. Despite a patient's blood glucose being low, this mass will continue to secrete insulin leading to dangerously low blood glucose levels that can be life-threatening. If not identified early on, this can lead to coma, seizures and ultimately death. It is a rare disease, and given that, this is a lower differential on a physician's diagnosis when a patient presents symptomatically. In the setting of a young patient exhibiting a behavioral disorder, trance-like states and disorientation perhaps epilepsy, alcohol withdrawal, drug use or drug withdrawal, psychiatric illnesses are more likely differential diagnoses than compared with a rare diagnosis of an insulinoma. Being very rare, it is not a disease that is seen as much as other diseases and perhaps not thought of early on. During the point in time of evaluation in an outpatient setting with non- fasting labs or an emergency department after patient had eaten, the blood glucose levels tested in the normal range can throw off the initial diagnosis. Complete surgical resection of the tumor is the definitive treatment [2].

Case Report

A 21 year old Caucasian male with a past medical history of MEN1 syndrome presents to the emergency department with a new seizure. No history of illicit drug use. Drinks alcohol occasionally. Family history is pertinent for father with MEN1 syndrome as well as two siblings with MEN1 syndrome. Prior to the seizure he was having 2-3 weeks of intermittent odd behavior, staring "trance – like" episodes and disorientation. One of these episodes occurred when he came home from work "in a daze" and symptoms resolved after eating cereal. His mother reported an instance where he returned home from work and acted disoriented and "odd". The patient ate and returned to his normal self. His mother took him to their primary care physician who referred him an outpatient Neurologist.

On the night of December 18, 2019, patient recalled turning off Christmas lights at home and going to sit on the couch. After which he did not recall anything else. His parents were woken up by their family dog barking. They then saw him having a whole body seizure. Patient did not have bladder or bowel incontinence or tongue biting during the episode. In the emergency department his blood glucose was noted to be 29. Urine drug screen was negative. Head CT was unremarkable. Neurology was consulted. He was started on a continuous infusion of dextrose IVF. Patient was transferred to our Cancer Center.

He was unable to undergo a 72 hour fast due to dangerously low levels of blood glucose within that timeframe. Was placed on a continuous dextrose IVF drip and started on diazoxide to help decrease insulin secretion. Laboratory work-up revealed an increased C peptide level of 2.6 and increased insulin level of 14. His IGF-1 and pro-insulin levels were elevated. PTH intact 67.2, 25-hydroxyvitamin D2 and D3, total low at 19.2, a high calcium of 12.0, 25-Hydroxyvitamin D2 less than 1.0, 25 -Hydroxyvitamin D3 19.2, 1,25 vitamin D 55.5.

CT abdomen and pelvis revealed masses in both the tail and body of the pancreas. See figure 1a and figure 1b. Further work up with an MRI of his abdomen showed similar findings. See figure 2a and 2b. He underwent an endoscopic ultrasound (EUS) that showed hypoechoic masses in the pancreas. He subsequently underwent a robotic assisted distal pancreatectomy and splenectomy. Tissue biopsy demonstrated tumor cells positive for insulin. On outpatient follow-up his blood glucose readings remained in normal ranges with resolution of his symptoms and no hypoglycemic episodes reported.

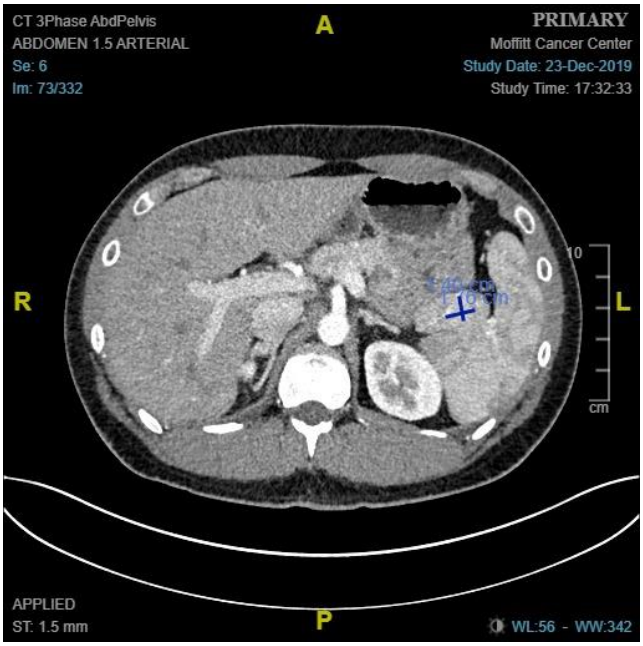

Figure 1a: Within the pancreatic tail is a 1.4 x 1.8 x 1.4 cm arterial enhancing lesion. It appears confined to the pancreatic parenchyma.

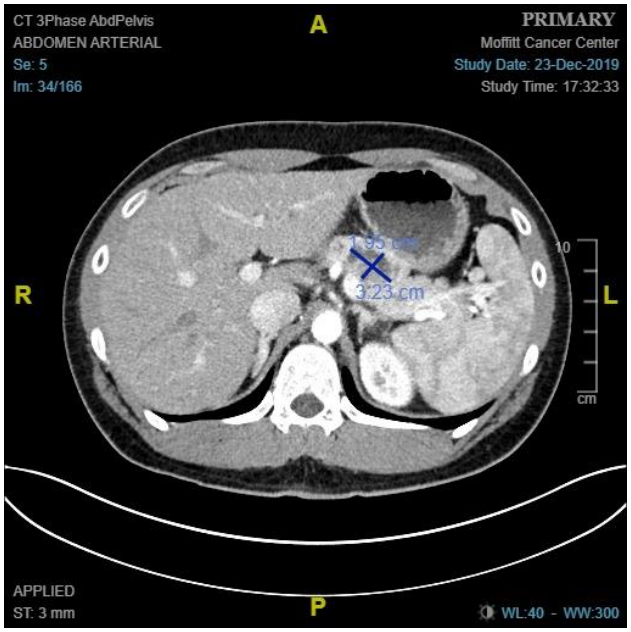

Figure 1b: Within the pancreatic body is a partially cystic mass that measures 1.95 x 3.23 x 2.8 cm.

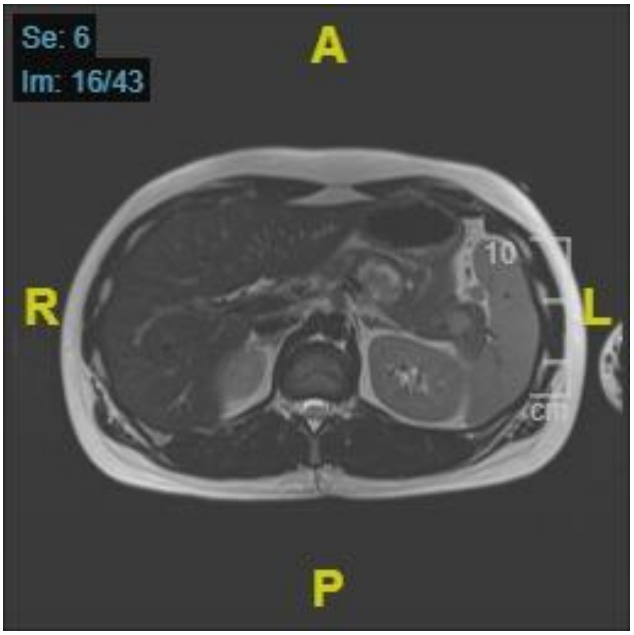

Figure 2a: Homogenously enhancing lesion in the distal tail of the pancreas measuring 1.4 cm.

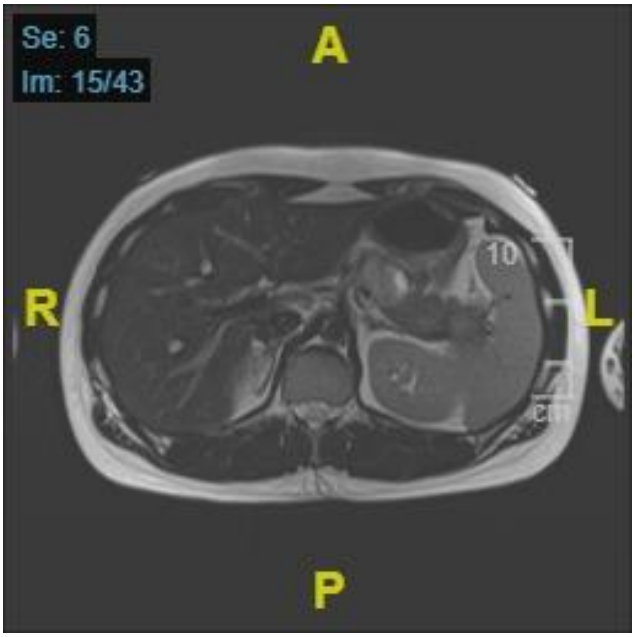

Figure 2b: The pancreas demonstrates a heterogenous complex lesion in the proximal tail measuring 3.3 x 2.3 cm.

Discussion

This study reports a case of insulinoma in a young adult presenting as "daze-like" states of intermittent confusion and odd behavior. These personality changes can be mistaken for other more common neuropsychiatric disorders including stroke, epilepsy, adrenal insufficiency, drug use, alcoholism. There is a broad differential spectrum of possible causes of this patient's symptoms. The key difference in this disease is the blood sugar. In insulinoma, we see life threatening levels of hypoglycemia [3]. Sometimes this can be missed on lab work since it occurs in an intermittent pattern.

The diagnosis typically requires continuous glucose monitoring along with lab work meeting the following criteria: prolonged fasting with serum insulin of ≥ 6 μU/ml, C-peptide ≥ 0.2 nmol/l and proinsulin ≥ 5 pmol/l [2]. Patient usually undergoes a 72 hour fast with continuous monitoring of glucose and usually stopped when glucose levels reach under 50 mg/dl. Patients with insulinoma may not make it to 72 hours due to profoundly low blood glucose levels during testing [4].

Imaging modalities includes but not limited to transabdominal ultrasound, CT, MRI, pancreatic arteriography, endoscopic ultrasound [3]. Pancreatic arteriography now has lower success rates of identifying disease than previously thought [5]. Localization of disease ranges from 25%- 50% with pancreatic arteriography. Transabdominal ultrasound can localize anywhere from 9%-66%. MRI identifies insulinomas in range of 40%-70%. CT 50%- 80%. EUS tends to have a higher range of sensitivity up to 93% [5].

Surgical management is the gold standard treatment of choice [6]. This provides a cure rate of 90 % [7]. These tumors are usually encapsulated making surgical excision precise [8]. Symptoms typically resolve post-operatively and patients can lead a normal life. If surgery is not an option or if a patient has episodes of severe hypoglycemia prior to the operating room, there are options for medical management. Medical management includes diazoxide, octreotide and lantreotide [4]. Diazoxide stops insulin release from pancreatic β cells [5]. The somatostatin analogues (octreotide and lantreotide) lower insulin levels in the plasma. Patients are instructed to eat frequent meals throughout the day to avoid hypoglycemia.

Conclusion

Insulinoma is a very rare tumor presenting with an incidence of 4 per 1 million persons per year. It can present with neurologic symptoms related to profound hypoglycemia. It is a disease that is less thought of as a differential diagnosis due to its rare nature. Patients can have seizures, become confused and disoriented. It is imperative to remember insulinoma as a cause of unexplained behaviors in a patient to avoid permanent neurologic damage. By the time a patient seeks evaluation, blood glucose may be normal and the diagnosis missed. Therefore, it is recommended that a patient be worked up with a 72-hour fast while continuously monitoring blood glucose. Rates of recurrence are extremely low (3%) and patients can enjoy a normal life after surgical treatment [5].

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