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Case Report

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Intractable Seizures an Abdominal Diagnosis

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1. Summary

Multiple endocrine neoplasia (MEN-1) is a rare Autosomal dominant disorder usually presenting with tumours of parathyroid gland, anterior pituitary and pancreatic islet cells [1]. Insulinoma is a rare neuroendocrine tumour originating from the beta cells of islets of Langerhans in the pancreas. It is the most common occurring neuroendocrine tumour after a non-functioning neuroendocrine tumour. However, with multiple endocrine neoplasia (MEN-1) syndrome, gastrinoma is more common. We share a rare case of insulinoma associated with MEN 1 syndrome with a rarer presentation, detected on dotanoc PET.

2. Background

Multiple endocrine neoplasia is a rare disorder and usually presents with hyperparathyroidism features as more than ninety percent of times parathyroid adenoma is present which is multifocal. The occurrence of neuroendocrine tumour is about fifty to sixty percent with most associated tumour being gastrinoma. A gentleman in his 6th decade presented with intractable seizures. On thorough evaluation he was detected of having insulinoma associated with MEN 1 syndrome.

3. Case Presentation

A gentleman in his mid-fifties presented with intractable seizures for 6 months. He was evaluated by a neurologist and was advised antiepileptics and was monitored. He again developed another episode of seizures, two months after the first episode and was managed with medication. He was evaluated with electroencephalogram and computed tomography brain which were normal. Random sugar levels were very low and was started on intravenous dextrose and referred to surgical gastroenterology and endocrine department for further management.

4. Investigations

Random blood sugar levels were measured regularly which showed values less than 50mg/dl, patient showing neuroglycopenic symptoms and would resolve after 25% dextrose infusion. This triad and its manifestations and corrections were observed for two days. Rest all routine investigations were in the normal range. However serum calcium was elevated13.0mg/dl (normal range 8.8-10.6mg/dl), S Parathormone was 222 pg/ml (normal range- 15-25pg/ml) , fasting insulin was 71.4mIU/L (normal range < 25mIU/L), C-peptide-6.63ng/ml (normal range- 0.5-2.7 ng/ml), random blood sugar- 36mg/dl. Serum cortisol was normal. CECT abdomen was done which showed a hyper-enhancing lesion in the pancreatic body 3x3cm size was noted. Hence, he was subjected to gallium dotanoc scan which showed two lesions, one as the previous one and a small lesion 0.6x0.5 cm in the distal body of pancreas along with an adrenal adenoma in the left adrenal gland and a parathyroid adenoma in the left side. There was no evidence of distant metastasis or any pulmonary nodules.

5. Differential Diagnosis if Relevant

With the above-mentioned presentation, the initial diagnosis was insulinoma as the whipple's tirad was visible. However, after all the blood investigations it was found to be Multiple endocrine neoplasia associated with insulinoma which is rare since most common tumour associated with Multiple endocrine neoplasia 1 is gastrinoma.

6. Treatment if Relevant

Since patient had life threatening episode of hypoglycaemia, he was planned for upfront pancreatic surgery, deciding to deal with para-

thyroid and adrenal adenoma at a later date.Initially spleen preserving distal pancreatectomy was planned. However, due to a large pancreatic tumour (3cm) with another lesion being intrapancreatic and splenic vein buried within the pancreatic parenchyma in addition to the malignant potential of the insulinoma associated with MEN-1 syndrome, spleen could not be preserved and the patient underwent distal pancreatosplenectomy.

7. Outcome and Follow-Up

Post operative period there were no further episodes of seizures or loss of consciousness. However, patient developed hyperglycaemia in the post operative period which was managed with regular management of sugar levels and insulin with the help of endocrinology department. On post operative day 4 patient developed a controlled grade A Pancreatic fistula which was managed conservatively. He was discharged on postoperative day 11 and was further sent to endocrinology for counselling and genetic testing. Histopathological report was suggestive of two neuroendocrine tumours with mitotic rate of 1/10 high power field with ki 67 <3% and no perineural and lympho-vascular invasion. Final diagnosis was Multiple endocrine neoplasia-1 Syndrome with insulinoma (pT2N0) stage IIb.

8. Discussion

MEN1 is diagnosed when two or more MEN1- associated endocrine tumors are present [1]. Parathyroid adenoma resulting in primary hyperparathyroidism is the most common feature of MEN1 and occurs in approximately 95% of MEN1 patients [2]. It is the most common presentation of MEN-1 syndrome [3]. Goudet et al. showed that first symptoms were related to hyperparathyroidism in 75% and insulinoma in 12% of the patients [4]. 10-60% of patients with MEN1 have pituitary tumors being prolactinoma the commonest tumor and the majorities are microdenoma [5]. Insulinomas are typically sporadic, benign tumours, with a solitary small (<2 cm in diameter) mass [6]. However, 10% of insulinomas are multiple, small in size and occur as a part of MEN type one syndrome. The diagnosis is often delayed or missed due to the rarity of insulinomas and nonspecific symptoms. Most symptomatic patients present with hypoglycemic episodes resulting from inappropriate intermittent insulin secretion by the tumour. Diagnosis is achieved by satisfying the criteria of Whipple's triad—hypoglycemia (plasma glucose <50 mg/dL), neuroglycopenic symptoms, and prompt relief of symptoms following the administration of glucose. The gold standard for biochemical diagnosis includes measuring plasma glucose, insulin, C-peptide, and proinsulin during a 72-hour fast. This prolonged fasting test can detect up to 99% of insulinomas. Approximately 65% of patients will experience hypoglycemic episodes within the first 24 hours of fast [7]. Radiological investigation usually contrast enhanced computed tomography is done which might help in localizing the tumour in arterial phase due to its hyperenhancement. Dotanoc scan is another scan which helps in localizing pancreatic neuronendocrine tumours, however not very sensitive for insulinoma. If it is not localized through the scans, Somatostatin receptor scintigraphy can also be done however due to paucity of somatostatin receptors in insulinoma it is not very sensitive. Intraoperative ultrasound is very useful in conditions were radiological localization failed but clinical manifestations are present (Figure 1-5).

However, our patient presented with intractable seizures which was initially deemed as neurological problem and later on thorough evaluation was found to have an insulinoma with multiple endocrine neoplasia-1. Moreso, the size of the tumour was unusually large (3cm) compared to usual sizes(<2cm) of multifocal insulinoma associated with MEN 1 syndrome. Insulinomas typically go undetected on dotanoc scan, however in this case both the lesions were dotanoc avid. With MEN-1 syndrome the most common neuroendocrine tumour associated is Gastrinoma however, with this case it was insulinoma. Initially planned for a spleen preserving distal pancreatectomy, however due to the large lesion involving the hilum of the spleen and the association of MEN-1 syndrome which adds up to the malignant potential of the insulinomas, open distal pancreatosplenectomy was done.

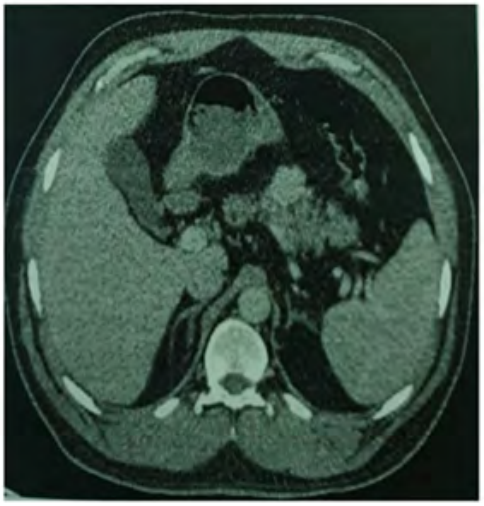

Figure 1: CECT abdomen shows a lesion in the body of the pancreas about 3x3 cm with hyperenhancement in the arterial phase.

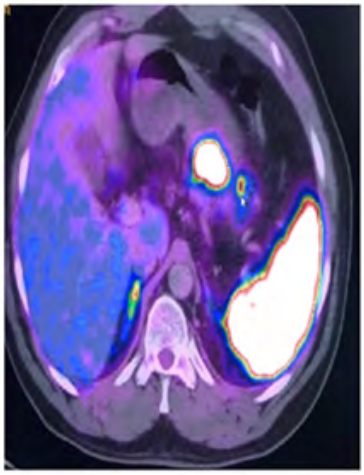

Figure 2: shows Gallium DOTANOC PET scan showing 2 lesions, one in the body 3x3 cm and another one 0.6x0.5 cm in distal body of pancreas

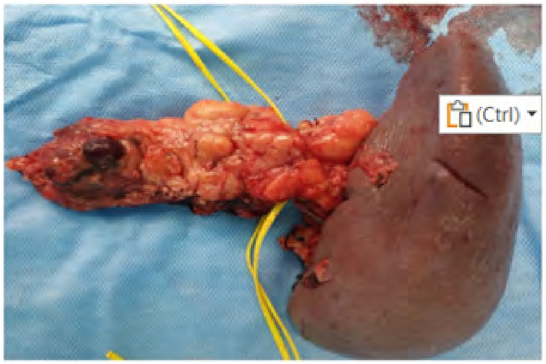

Figure 3: Resected specimen photo with distal pancreas, spleen and reddish-brown tumour in the body region.

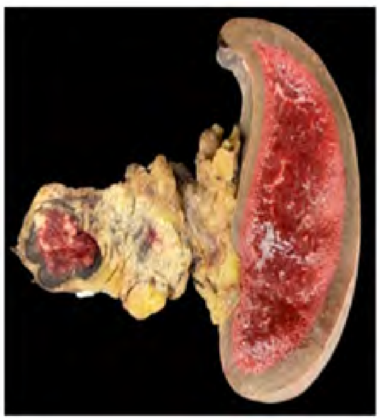

Figure 4: Shows cut section of the specimen with both tumours visible in the body and distal body of the pancreas.

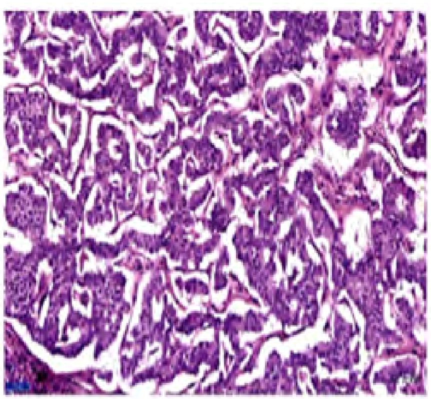

Figure 5: Salt and pepper chromatin with trabecular pattern and hyalinized stroma

9. Learning Points/Take Home Messages 3-5 Bullet Points

•Multiple endocrine neoplasia-1 is a rare syndrome.

•It usually presents with hyperparathyroidism features, however our patient presented with an atypical presentation with intractable seizures.

•Usually insulinomas with MEN-1 are small in size, however here it was large in size.

•Insulinomas usually associated with Multiple endocrine neoplasia 1 are associated with malignancy.

•Insulinomas are usually not detected on Dotanoc scan due to absence of somatostatin receptors, however this tumour was evident on dotanoc scan.

•Hence, it should be remembered that atypical presentations may occur.

10. Patient's Perspective

I am thankful to the surgical team for detection of my disease and uneventful surgery and early recovery.

11. Declaration of Conflicting Interests

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