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Case Report
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Case Report: Pseudo-Mirizzi Syndrome as a Rare Initial Clinical Presentation of MEN1 Syndrome

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Abstract

Introduction: Pseudo Mirizzi Syndrome (PMS) refers to extrinsic compression of the bile duct without calculi. PMS due to metastatic lymph node in Multiple Endocrine Neoplasia type 1 (MEN1) has not previously been described, to our knowledge, this is the first reported case of PMS in MEN 1 syndrome.

Case Presentation: A 48-year female presented with upper abdominal pain and fever. Examination revealed tenderness in the right upper abdomen; the rest of the abdomen was unremarkable. Ultrasound and MR imaging suggested acute cholecystitis with features of Mirizzi syndrome. Laparoscopic cholecystectomy was performed due to persistent pain and ultrasound evidence of Gallbladder (GB) perforation. Intraoperatively, the GB was distended, and a large pericholedochal lymph node was found compressing the mid common hepatic duct. Histopathology confirmed necrotizing cholecystitis, and the lymph node showed metastatic, Neuroendocrine Tumor (NET). Further evaluation revealed a Ga-68 DOTANOC avid pancreatic and liver lesion, pituitary macroadenoma and parathyroid adenomas, consistent with MEN1 diagnosis. Genetic testing confirmed a pathogenic MEN1 mutation.

Conclusion: This report underscores the importance of recognizing atypical causes of biliary obstruction, such as Pseudo-Mirizzi syndrome due to metastatic lymph node involvement, particularly in patients with syndromic conditions like MEN1. Clinicians should maintain a high index of suspicion when encountering unusual biliary manifestations.

Keywords: Acute cholecystitis; Gastrinoma; MEN1 syndrome; Pancreatic neuroendocrine tumor; Pituitary macroadenoma; Pseudo mirizzi syndrome

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Introduction

Pseudo-Mirizzi Syndrome (PMS) is a rare variant of Mirizzi syndrome, where biliary obstruction occurs due to extrinsic compression rather than a cystic duct stone. Only few cases of PMS are reported in English literature. In previously reported PMS cases common hepatic duct obstructed by edematous gall bladder [1,2]. In our case, the obstructive element was a pericholedochal lymph node harboring a metastatic NET. Multiple Endocrine Neoplasia Type 1 (MEN1) is an autosomal dominant syndrome characterized by neoplasms primarily affecting the parathyroid glands, pancreatic islets, and anterior pituitary. Clinical manifestations vary according to tumor type and hormone secretion [3]. We report a rare initial clinical case presentation of MEN1 as Pseudo-Mirizzi syndrome with acute cholecystitis.

Case Presentation

A 48-year-old obese woman (BMI 28.5 kg/m^2) presented with right upper abdominal pain and fever. Her medical and personal history was unremarkable. Notably, her brother had previously undergone brain surgery for a pituitary adenoma. On examination, she had tenderness in the right upper abdomen, with no other abnormal findings. Cardiovascular, respiratory, and neurological systems examinations were normal. Blood tests showed leukocytosis (19,200/µL), serum alkaline phosphatase level was raised (250 U/L), rest of liver and kidney function tests were normal. Abdominal ultrasound revealed a distended, edematous gallbladder with a stone impacted

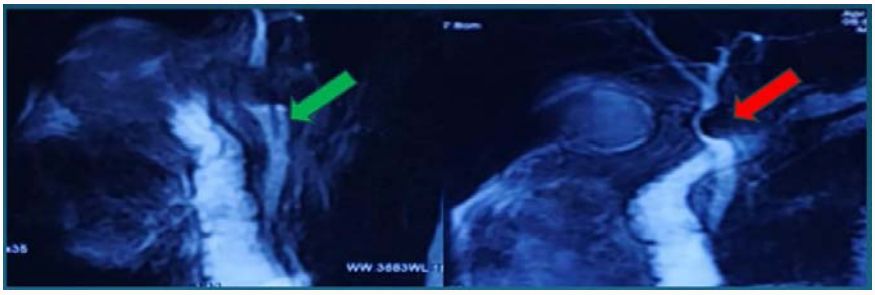

Figure 1: Magnetic Resonance Cholangiopancreatography (MRCP). Showing medial insertion of cystic duct (Green arrow) with filling defect (Red arrow) in it which is compressing over CBD.

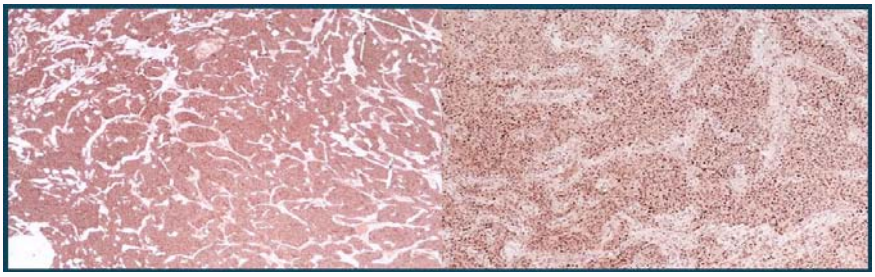

Figure 2: Histopathology report. LN showing tumor cells stained positive for synaptophysin (left) and INSM1 (right).

at the neck, compressing the Common Hepatic Duct (CHD). MRCP confirmed gallbladder distension with a stone at the neck compressing the CHD with proximal CHD dilatation, suggestive of Mirizzi syndrome (Figure 1).

She was admitted and started on conservative treatment for acute cholecystitis (Tokyo grade II) with suspected Mirizzi syndrome. By day six, her pain and fever increased with leukocytosis, prompting laparoscopic surgery. Intraoperatively, the gallbladder was edematous with pericholecystic fluid and dense omental adhesions (Parkland Grade III cholecystitis). The Calot's triangle defined. No stones were found in the cystic duct, but a large lymph node was noted compressing over the CHD. Cholecystectomy with lymph node sampling was performed. These intraoperative findings were consistent with PMS, as the CHD compression was due to the lymph node rather than the stone. She recovered uneventfully and was discharged on postoperative day five. Gallbladder histopathology showed necrotizing cholecystitis. The lymph node biopsy revealed metastatic, well-differentiated Neuroendocrine Tumor (NET). Immunohistochemistry was positive for chromogranin and synaptophysin (Figure 2).

Following this unexpected finding, a Gallium-68 DOTANOC PET-CT (Figure 3) demonstrated Somatostatin Receptor (SSTR) expression in the pancreatic body, periampullary region, and liver, consistent with metastatic Pancreatic NET (PNET). Given the scan results and her family history of pituitary tumor, evaluation for multiple endocrine neoplasia type 1 (MEN1) was initiated. Neck ultrasound identified parathyroid adenomas, and brain MRI (Figure 4) revealed a 1.5 × 1.6 cm pituitary macroadenoma. Both were asymptomatic. Serum chromogranin A was elevated at 300 ng/mL. PTH was 100 pg/mL; anterior pituitary hormones were within normal limits. Genetic testing confirmed a pathogenic mutation in exons 2-10 of the MEN1 gene, confirming MEN1 syndrome.

She was started on long-acting octreotide (30 mg, monthly) for

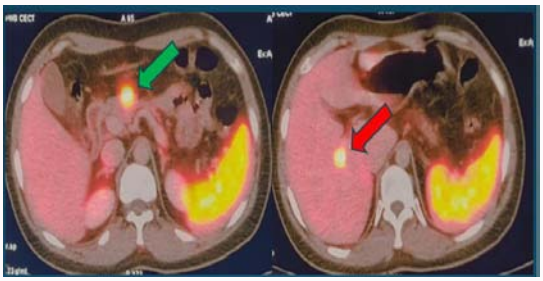

Figure 3: DOTANOC Scan. DOTANOC Scan showing increased SSTR expression in pancreatic head region (green arrow) and liver (red arrow; suggestive of metastasis).

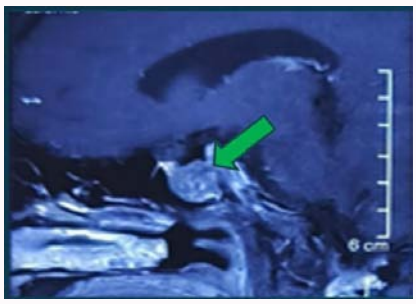

Figure 4: MRI Brain. MRI Brain showing ~2 cm sized lesion in Sella turcica suggestive of pituitary macroadenoma.

metastatic PNET. During follow-up, she developed hypercalcemia (serum calcium 13.6 mg/dL) and blurred vision. Due to symptomatic hyperparathyroidism and the pituitary adenoma, she was scheduled for staged surgery. Tc-99m MIBI scan revealed bilateral inferior parathyroid adenomas. She underwent subtotal parathyroidectomy (removal of 3½ glands), followed by transsphenoidal resection of the pituitary macroadenoma.

One year later, she reported epigastric burning, relieved with proton Pump Inhibitors (PPIs). Upper GI endoscopy revealed sessile nodules along the greater curvature of the stomach. Biopsy confirmed WHO Grade 1 NET, positive for chromogranin and synaptophysin. After discontinuing PPIs for 14 days, serum gastrin was measured at 1200 pg/mL, confirming a diagnosis of gastrinoma.

Currently, she remains clinically stable on somatostatin analog therapy for metastatic PNET and continues PPI therapy for gastrinoma. Her calcium levels are normalized, and follow-up DOTANOC scans show stable disease. Hormonal panels are monitored routinely, and her family, has received genetic counseling.

Discussion

MEN1 was described by Paul Wermer in 1954 as an autosomal dominant disorder. It most commonly targets the parathyroids, pancreatic islet cells, and anterior pituitary. The estimated prevalence is about 1 in 30,000 individuals, with no known gender or ethnic bias [1]. Diagnosis is usually made when two of the three principal tumors are present, though as a rare presentation thymic and lung NETs, adrenocortical lesions, lipomas, angiofibromas, and collagenomas have been documented [2,3].

Our case presented with a highly unusual form of biliary obstruction Pseudo-Mirizzi syndrome which led to the diagnosis of MEN1. PMS involves extrinsic compression of the bile duct without stones, in contrast to classical Mirizzi syndrome. While most PMS cases [4,5] are attributed to an inflamed or distended gallbladder, ours involved a metastatic neuroendocrine tumor infiltrating a pericholedochal lymph node, exerting pressure on CHD. This represents, to our knowledge, the first documented instance of PMS in MEN1.

The patient also had Grade II acute cholecystitis, managed according the Tokyo Guidelines 2018 [6], which recommend early surgery for moderate disease. Intraoperative findings showed distended gallbladder with omental adhesions and pericholecystic fluid, consistent with Parkland Grading Scale (PGS), difficulty grade III [7]. Calot's triangle was successfully exposed after adhesiolysis, and cholecystectomy was completed.

Typically, MEN1 presents with common clinical presentation. MEN1-related primary hyperparathyroidism typically involves all four glands. Most cases are managed surgically with subtotal parathyroidectomy (3-3.5 glands) plus thymectomy. Even after surgery, recurrence is frequent, and calcimimetics or ethanol ablation may be used in selected cases [3,8].

Pancreatic and duodenal neuroendocrine tumors (dpNETs) affect over 80% of MEN1 patients and are the leading cause of disease-related mortality. These may be functioning (e.g., insulinomas, gastrinomas) or non-functioning; the latter are often small and asymptomatic. Current guidelines recommend surgical removal of all functioning tumors and any non-functioning lesions ≥2 cm or lesion showing growth. For metastatic or symptomatic disease, treatment options include somatostatin analogues, liver-directed therapy, or PRRT with ^{177}Lu-DOTATATE [3,8].

Lastly, pituitary adenomas occur in approximately 40% - 50% of MEN1 patients and may occasionally be the initial presentation, with prolactinomas being the most common subtype. Diagnosis relies on hormone assays and contrast-enhanced MRI. Cabergoline is the first-line medical therapy for prolactinomas, while surgical management is reserved for large, unresponsive, or symptomatic lesions [3,8].

Conclusion

This report underscores the importance of recognizing atypical causes of biliary obstruction, such as Pseudo-Mirizzi syndrome due to metastatic lymph node involvement, particularly in patients with syndromic conditions like MEN1. Early identification of such rare presentations can lead to timely diagnosis and management of MEN1. Clinicians should maintain a high index of suspicion when encountering unusual biliary manifestations.

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