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A Rare Case of Duodenal Somatostatinoma

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Abstract
Somatostatinomas are the rarest of pancreatic tumours and can arise in pancreas and duodenum.Duodenal somatostatinomas are less common and are distinguished from their pancreatic counterparts by a frequent association with type I neurofibromatosis, the presence of psammoma bodies,the less frequent presence of metastatic disease and absence of somatostatinoma syndrome.
We present a case of somatostatinoma in a 70 year old woman who presented with vague epigastric pain and nausea. In view of CT and MRCP finding suggestive of malignant lesion in periampullary region, patient was posted for exploratory laparotomy and Whipple's procedure was done. Histopathology report revealed a somatostinoma. Postoperative course was uneventful and patient was discharged after 15 days. After 2 years of follow up patient was disease free.

INTRODUCTION

Somatostatinoma,arising from delta cells in the pancreas ,is a rare tumor of the gastroenteropancreatic neuroendocrine axis,representing G-I of gastrointestinalneuroendocrine tumors (NET). The first case of somatostatin secreting tumors were reported in 1977, after which fewer than 200 cases were reported . about 2/3 rd of these tumors arise in pancreas ,with remainder arising in the duodenum, with rare primary sites including liver,colon and rectum. The majority of the tumors are malignant with variation in the frequency with

which somatostinomas secret somatostatin. Some somatostatinomas particularly those arising in the ampullary and periampullary region are not associated with any functional syndrome,with only 10% of the patients experiencing somatostatinoma syndrome. Most common clinical presentation is abdominal pain with nausea and weight loss. We present duodenal somatostatinoma and review gastrointestinal somatostatinomas.

CASE REPORT

A 70 yrs old female patient presented to our hospital with complaints of vague epigastric pain and nausea for 15 days . abdominal examination revealed no abnormality. Her total serum bilirubin was 0.9 mg/dl and SGOT/SGPT were 10/20 IU/L, with other laboratory findings being normal. The tumor markers CEA and CA 19-9 were in normal range. There were no signs of neurofibromatosis . ultrasound showed a dilated CBD (15mms) with bulky head of pancreas near ampulla.CT scan showed minimally enhancing mass lesion measuring approximately 7mms near ampulla of vater causing obstruction of both CBD and pancreatic duct(fig 1). MRCP showed obstruction of terminal portion of CBD with dilated CBD and pancreatic duct.(fig 2)

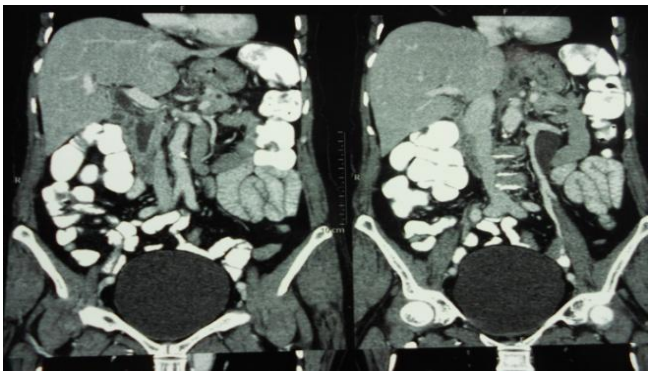

Fig 1: CT scan showing bulky pancreas with minimally enhancing mass lesion measuring approximately 7mms near ampulla of vater causing obstruction of both CBD and pancreatic duct

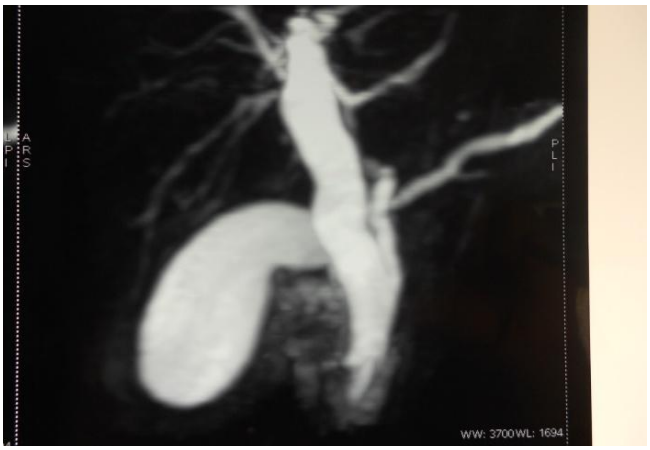

Figure 2: MRCP showing obstruction of terminal portion of CBD with dilated CBD and pancreatic duct

In order to resolve the diagnostic dilemma, we decided to explore the patient. Intraoperatively a hard mass arising suspicion of malignancy was palpable in the 2nd part of duodenum. In view of the above findings decision to proceed with Whipples operation was taken.

Macroscopically , cut section showed ill defined mass in 2nd part of duodenum causing flattening of mucosal folds and partial luminal compromise,

with infiltration of surrounding pancreatic parenchyma.(fig 3)

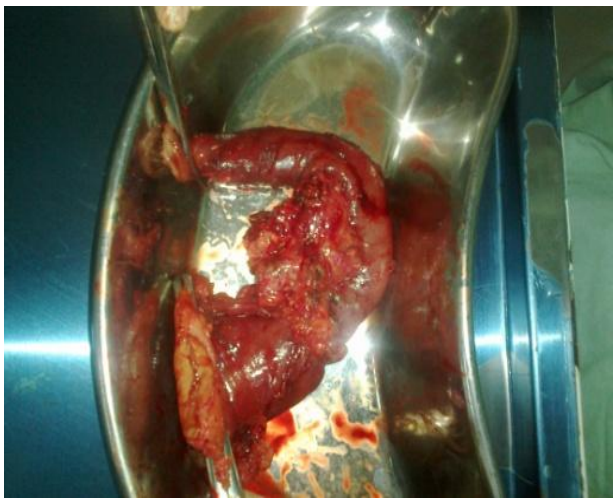

Figure 3:Specimen after whipples surgery.

Histopathology showed tumor invading submucosa and muscle coat of duodenum with focal involvement of duodenum with focal involvement of overlying mucosa. It consisted of uniform cells arranged in nests ,cords and glandular patterns with psammoma bodies. All resected tumor margins and lymphnodes were free of tumor. The tumor cells stained posive for CK-7 and Neuron specific enolase and are negative for CK 20 ,synaptomycin and chromogranin.(fig 4, 5)

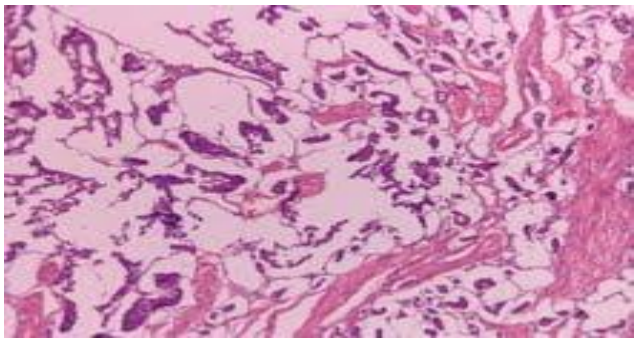

Figure 4: Histopathology showing tumor invading submucosa and muscle coat of duodenum with uniform cells arranged in nests ,cords and glandular patterns with psammoma bodies.

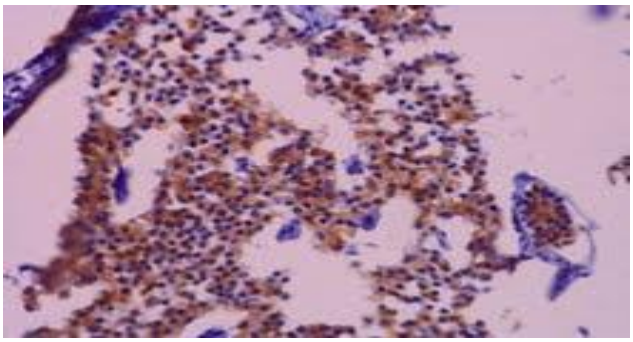

Figure 5:IHC stained posive for CK-7 and Neuron specific enolase and are negative for CK 20 ,synaptomycin and chromogranin

We diagnosed it as duodenal somatostatinoma.

Postoperative course was uneventful and patient was discharged after 15 days. 2 years follow up was unremarkable.

DISCUSSION

Somatostatinomas are extremely rare of the commonly accepted functional APUD(Amine Precursor Uptake and Decarboxylation)cell neuroendocrine tumors of Gastrointestinal Tract with an annual incidence of 1 case per 40 million in adults.In 1977,Larsson et al , independently reported the first case of somatostatinoma which was followed by full description of somatostatinoma syndrome in 1977 by Krejs and colleagues.(1,2) Somatostatinoma is a tetradecapeptide that inhibits numerous endocrine and exocrine secretory hormones including insulin,cholecystikinin,pancreatic enzymes and gastrin.It has marked effects on Gastrointestinal transit time,intestinal motility and absorption of nutrients.

Approximately,one half of these tumors are in the pancreas,two-thirds of which arise within the head

of the gland.The remainder arise largely in the duodenum,particularly in the ampullary and periampullary region.The mean age at diagnosis is 50 years(range-26 to 84) with a roughly equal distribution between men and women.

The most common presenting symptom of a patient with somatostatinoma is abdominal discomfort. Pancreatic tumors tend to be greater than 4 cm while most duodenal tumors are less than 2cm at diagnosis.As a result of their location,patients with duodenal tumors often present earlier with bile duct obstruction and jaundice,hence the reason for their early discovery,small size and low rate of metastasis.Unlike,insulinomas and gastrinoma which tend to become symptomatic due to hormone overproduction at a small size,symptoms of somatostatinoma due to hormone excess occurs with high hormone levels and with large tumors.The reported incidence of "Somatostatinoma syndrome" is 10% which involves gallstones,diarrhea ,steatorrhoea and mild diabetes.About 50% of somatostatinomas have other neuroendocrine disorders,particularly those presenting with duodenal tumors and include NF-1 and less frequently MEN-1,V.H.L.disease,Tuberous Sclerosis and G.I.S.T.It is vital to confirm or refute these associated conditions.

About 2/3rds of duodenal somatostatinomas contain psammoma bodies within the lumen of their glandular structure.Tumor markers with exception of Somatostatin are non specific and include Pancreatic Polypeptide,Ghrelin,Neuron Specific Enolase and Chromgranin A.

The preoperative diagnosis of somatostatinoma is extremely difficult to establish unless there is a high clinical suspicion based on the patient's symptomatology, although elevated fasting serum levels of somatostatin that cause the somatostatinoma syndrome can be diagnostic, particularly if associated with tumors larger than 4 cm. CT and magnetic resonance imaging often fail to identify small tumors of the duodenum, and these are often discovered during upper gastrointestinal endoscopy. The diagnosis is then made based upon histological assessment of biopsy specimens during upper gastrointestinal endoscopy or endoscopic US; with the exact histopathological diagnosis ultimately based on the resected specimen Surgical resection is the only form of curative treatment smaller lesions (< 2 cm) by enucleation larger lesions with extensive Whipple's type resection. (due to their common localization in the head of the pancreas and duodenum), or other similar anastomoses. Unfortunately, in only 60%-70% of surgical cases is the tumor completely resected. Debulking surgery should also be considered in the presence of metastatic secondaries.

For patients with liver secondaries that are not amenable to resection or ablation, hepatic artery embolization or chemoembolization may be an effective therapy for symptomatic palliation. Adjuvant chemotherapy is not advocated after complete resection, however, in locally unresectable cases or in metastatic

somatostatinoma, chemotherapeutic agents have been used with moderate clinical responses.

The overall 5-year survival for patients with somatostatinoma is 40% to 60%. The 5-year survival rate is 40% in somatostatinomas with liver metastasis, but 100% in tumors without liver or lymph node metastases.

Ideal Treatment for this patient is whipples procedure but there has been case reports of local excision of mass from duodenum with sphincteroplasty(3). In present case we suspected malignant tumor by intra operative palpation of hard mass and decided for pancreatoduodenectomy. Even Though this patient was 70 year old with high risk for major surgery, she recovered well post operatively and is symptom free till this day. After histopathological examination and immune histochemistry it was proved to be somatostatinoma. On retrograde analysis of symptoms of this patient we confirmed it as a non-secreting duodenal somatostatinoma. Due to absence of typical triad of cholelithiasis, steatorrhoea and diabeties mellitus it is essential to have a high grade of suspicion for somatostatinoma and decide for radical surgical treatment to prevent high chance of early metastasis as it is a tumour with grave prognosis.

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