LOCAL RESECTION OF AMPULLARY NEUROENDOCRINE TUMOR WITH HIGH RISK FACTOR (FACTOR VII DEFICIENCY)-case report

RAMESH N NATARAJAN
Department of Surgical Gastroenterology and Proctology,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract:
ABSTRACT Ampullary neuroendocrine tumors are extremely rare tumors with an incidence of less than 0.3% of gastrointestinal carcinoids and less than 2% of all tumors of ampullary region. Ampullary neuroendocrine tumors can present with jaundice, or nonspecific abdominal discomfort or gastrointestinal bleeding. Endoscopic biopsy is done for histopathological examination and to analyze the tumor markers in the tissue like Chromogranin A and Neuron specific enolase. Pancreaticoduodenectomy is often recommended as treatment of choice for ampullary neuroendocrine tumor less than 2 cm size or in high risk surgical candidates. Our patient had factor VII deficiency and A Negative blood group.

Keyword: ampulla, factor VII

INTRODUCTION:
Ampullary neuroendocrine tumors are extremely rare tumors with an incidence of less than 0.3% of gastrointestinal carcinoids and less than 2% of all tumors of ampullary region. Ampullary neuroendocrine tumors can present with jaundice, or nonspecific abdominal discomfort or gastrointestinal bleeding. Endoscopic biopsy is done for histopathological examination and to analyze the tumor markers in the tissue like Chromogranin A and Neuron specific enolase. Pancreaticoduodenectomy is often recommended as treatment of choice for tumor of any size with no distant spread. However, less invasive procedures like local excision and endoscopic resection have also been successfully attempted especially for ampullary neuroendocrine tumor less than 2 cm size or in high risk surgical candidates. Our patient had factor VII deficiency and A Negative blood group.
CASE PRESENTATION.
48 years old female admitted with H/O fever, chills, rigor, melena, vomiting on and off for 20 days. She is a known case of factor V11 deficiency with cholelithiasis, choledocholithiasis on regular treatment elsewhere in private hospitals for the past 5 years. Three months back she underwent ERCP for above illness, ERCP showed bulging ampulla, biopsy from which showed features of neuroendocrine tumor, CBD stenting was also done. Within a week she was readmitted with upper GI bleeding with Hb value of 2.9. Endoscopy showed bleeding from the previous biopsy site and was controlled with adrenaline injection. She had second episode of bleeding 1 months later, which was managed conservatively in same hospital. She had another episode after 20 days and was referred to our hospitals for further management. Blood investigation done which was consistent with factor V11 deficiency and A negative blood group. Investigation showed Hb -7.1, PT-22.5, INR-1.8, fibrinogen -473/dl(N-180-350), PTT:29.45 (N-31.5). MRCP Revealed IHBR dilatation, distended GB with 12 mm calculi, dilated CBD with multiple calculi. Upper GI Endoscopy showed bulky Ampulla with surface ulceration, causing partial obstruction of duodenum with no fresh bleeding. Biopsy showed well differentiated neuroendocrine tumor with tumor cells diffusely positive for synaptophysin, chromogranin. As the patient comes under high risk category with factor VII deficiency and had a rare blood group (A Negative), minimal surgical procedure was planned—surgery plan was transduodenal local resection of neuroendocrine ampullary tumor. We decided against endoscopic therapy because of tumor without affecting the entry site of bile duct and pancreatic duct was done. Cholecystectomy with CBD exploration and T tube drainage was also done. 4 units of FFP transfused 1 hour before surgery and inj FEIBA (Factor eight inhibitor bypassing activity) -20000IU BD was given for 3 days after surgery. T tube cholangiogram was done on 7th POD showed no residual stones or leak. Post-operative biopsy showed duodenal neuroendocrine tumor with infiltration of underlying muscularis propria with margin positivity. IHC study showed strong positivity for NSE (neuron specific enolase) and chromogranin.

FOLLOW UP:
T tube cholangiogram done after 6 weeks was found to be normal. T tube was removed and the patient was kept under regular follow up once in 3 months. No evidence of abdominal pain, jaundice or bleeding reported. Liver function test, complete blood count and renal function test in follow up period was normal. USG abdomen was normal. Upper GI Endoscopy showed no evidence of recurrent growth.
DISCUSSION:
Among the gastrointestinal and pancreatic NET, the ampulla of Vater represents an uncommon site for the disease. CT, MRI, ultrasound endoscopy and endoscopic retrograde cholangiopancreatography with biopsy are the main tools for assessing NET of the ampulla of Vater preoperatively.
Currently, several options are available to treat NET of the ampulla of Vater. Endoscopic local resection and surgical ampullectomy have been considered to be safe for small NET of the ampulla of Vater (less than 2 cm) [2] or in patients with severe comorbidities (2).

A major limitation of these procedures is the inability to address loco-regional lymph node metastases, possibly leading to an inadequate oncological operation and potentially jeopardizing the patient’s prognosis [4]. Pancreatoduodenectomy has been generally considered the procedure of choice for NET of the ampulla of Vater that is larger than 2 cm and for cases of neuroendocrine carcinomas (5). In a recent multi-institutional review of patients with resected small neuroendocrine pancreatic and periampullary NET, local resection had similar results to pancreaticoduodenectomy in terms of overall survival but was associated with significantly lower morbidity (6). However, this study addressed a selected group of patients (with tumors less than 3 cm and no involved lymph nodes or liver metastases) (6). Moreover, in experienced centers, the morbidity and mortality rates of the Tran’s duodenal local excisions of ampullary lesions are comparable to pancreaticoduodenectomy [7]. Transduodenal ampullectomy was first introduced by Halstead in 1899. Operative procedure included the Kocher maneuver, longitudinal duodenotomy, full thickness excision of the ampulla, and re-implantation of the common bile duct and the pancreatic duct into the duodenal wall.

But in our case since the patient is a high risk surgical candidate—factor V11 deficiency/A negative blood group. We did only local resection of the tumor without affecting the bile duct and the pancreatic duct entry, which can be very well appreciated in peroperative photograph. Usually it is indicated in Benign Tumour of ampulla and Neuroendocrine tumour of ampulla <3 cm. Ampullectomy is
Less morbid procedure. Less invasive procedures like local excision and endoscopic resections have also been successfully attempted, especially for ANET less than 2 cm size or in high risk surgical candidates (8,9,10) Advantages of this procedure is 1. Pancreticobiliary duodenal continuity maintained 2. No nodal dissection done. 3. Risk of margin positivity is possible compared to radical surgery. Radical surgery i.e. pancreaticoduodenectomy with lymphadenectomy should be the standard approach in most patients with NET of the Ampulla of vater because this procedure removes the entire potential tumour bearing tissue. Extensive debulking surgery should be considered in patient with hormonal hypersecretion even in the presence of advanced disease (extensive local or Distant diseases it has been shown to offer survival rates of up to 80% at 5 yrs.). (11). Our patient had successful local resection of her tumour and now she is under follow up for the past 11 months. She is disease free till now.

CONCLUSION:
Ampullary neuroendocrine tumours are rare GI tumours but nevertheless should be included in the differential list for recurrent abdominal pain of unknown cause with upper or lower GI bleeding Though pancreaticoduodenectomy has been standard of practice; there is increasing evidence that local resection could be an alternative treatment modality with much less morbidity in special situation like this i.e. factor V11 deficiency and A negative blood group.


