



A rare case report of multiple and large, small bowel neuroendocrine tumours in a 50 year old lady

Sarojini P Jadhav¹
Harsh R Mehta^{2*}
Sriranjani Iyer²

¹Professor and Head, Department of General Surgery, GMC Aurangabad, India

²Junior resident, Department of General Surgery, GMC Aurangabad, India

Abstract

Small intestinal neuroendocrine tumors are the most common type of neuroendocrine tumors and small bowel neoplasms. The clinical presentation varies from asymptomatic incidentally detected localized lesions to lethal metastatic disease with carcinoid syndrome.

A 50 year old lady presented with chief complaints of lump in upper abdomen associated with dull aching pain for 3 months. Cect scan was suggestive of a probable gastrointestinal stromal tumour for which patient underwent elective laparotomy. Postoperatively on the basis of histopathology a diagnosis of neuroendocrine tumor was made and confirmed by immunohistochemistry. Approximately 90% of duodenal NET's are not associated with a clinical syndrome. It is a rare diagnosis with incidence of 0.5/100,000.

Patients with well differentiated tumours diagnosed in early stage have a good prognosis with an 85% overall 5year survival rate.

Keywords

Neuroendocrine; Gastrointestinal ; Stromal; Duodenum

Introduction

Neuroendocrine tumors are a heterogeneous group of neoplasms arising from various anatomical sites with the majority originating from small intestine and pancreas.

They originate in the enterochromaffin cells of the aerodigestive tract.

The underlying pathogenic mechanisms have been poorly characterized due to relative rarity [1].

Small intestinal neuroendocrine tumors are the most common type of neuroendocrine tumors and small bowel neoplasms. The clinical presentation varies from asymptomatic incidentally detected localized lesions to lethal metastatic disease with carcinoid syndrome [2].

Functional neuroendocrine tumors are characterized by a clinical syndrome caused by excess hormonal secretion while non-functional neuroendocrine tumors are hormonally silent [3].

Intestinal neuroendocrine tumors have distinct features depending on their site of origin.

Neuroendocrine tumors originating in the duodenum are not well characterized. Many are discovered incidentally during endoscopy [4]. Unlike other midgut neuroendocrine tumors, they are not commonly associated with the typical carcinoid syndrome.

NET's usually have an indolent clinical course, but tumor behavior varies widely depending on 2017 WHO (World Health Organization) grading, based on Ki-67 proliferation index and mitotic count [5].

Case Report

A 50 year old lady presented with chief complaints of lump in upper abdomen associated with dull aching pain since 3 months.

There were no associated complaints of pain, fever, vomiting, constipation, diarrhoea, passage of blood in urine or stools, breathlessness, loss of weight or appetite and no complaint of other swellings on the body.

The Patient was not a known case of diabetes mellitus, hypertension, heart disease, asthma or any other major illnesses. The Patient is not a known alcoholic, smoker or drug addict. There is history of attaining menopause 5 years ago. Presently the patient bears two children.

Article Information

DOI: 10.31021/ijcmc.20203146

Article Type: Case Report

Journal Type: Open Access

Volume: 3 Issue: 4

Manuscript ID: IJCMC-3-146

Publisher: Boffin Access Limited

Received Date: 27 February 2020

Accepted Date: 06 March 2020

Published Date: 10 March 2020

*Corresponding author:

Harsh Mehta

Junior resident,

General surgery

GMC Aurangabad, India

E-mail: harshees81@gmail.com

Citation: Jadhav SP, Mehta HR, Iyer S.A Rare Case Report of Multiple and Large Small Bowel Neuroendocrine Tumours in a 50 Year Old Lady. Int J Clin Med Cases. 2020 Mar;3(4):146.

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On physical examination, the patient was vitally stable. Per abdomen examination was soft to touch, non-tender with no guarding or rigidity felt. A lump of 15x8cm was felt in left hypochondriac and epigastric region, firm in consistency, immobile, non-tender with no local rise in temperature and with a smooth overlying surface. Per vaginal and per rectal examination had no abnormal findings.

Routine lab investigations were within normal limits

The Patient was referred with a contrast enhanced computed tomography scan of the abdomen and pelvis which was suggestive of

1. A large, well-defined solid heterogeneously enhancing exophytic mass lesion of size 12x8cm containing non enhancing necrotic areas, arising from the third part of duodenum.
2. Two well defined nodular enhancing lesions of approximate size 2x1 cm and 3x1cm from the second part of duodenum.

Following above findings, CT was suggestive of a probable gastrointestinal stromal tumour with paraduodenal nodes (Figure 1,2).

An upper gastrointestinal endoscopy was performed which was within normal limits. USG guided biopsy or FNAC was not done due to the high vascularity of tumour.

Following investigations and anesthesia fitness, the patient was posted for an elective exploratory laparotomy.

Intraoperatively

1. There was evidence of a large 15x8x6cm exophytic growth from third and fourth part of duodenum, duodenojejunal flexure and proximal jejunal segment.
2. Another 4x2cm and 3x2cm growths were seen rising from the second part of duodenum.

Intraoperative decision was taken to perform Whipple's procedure.

Resection of the large tumor with associated jejunal segment was done with primary closure of proximal and distal duodenum.

Cholecystectomy was done with resection of pylorus, duodenum and part of pancreas followed by sequential pancreaticojejunostomy, choledochojejunostomy, gastrojejunostomy and feeding jejunostomy (Figure 3 and 4).

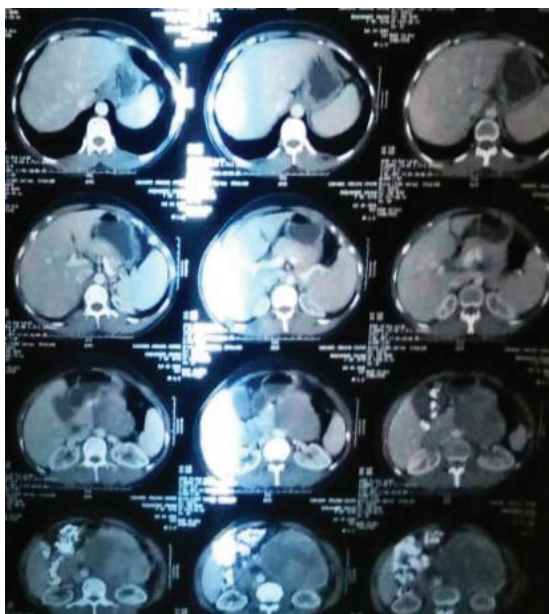


Figure 1: Shows extent of mass on CT scan



Figure 2: Showing extent and involvement of surrounding structures



Figure 3: Shows growth arising from duodenum and jejunum



Figure 4: Showing complete specimen after excision

Postoperatively the patient was managed aggressively and closely monitored. Multiple blood transfusions and parenteral nutrition were supplied consecutively. Feeding jejunostomy feeds were started on the fifth day postoperatively. Abdominal drain in subhepatic space was removed on the sixth day while the pelvic drain was removed on the tenth day postoperatively.

Histopathology report was as follows

1. Grade 1 neuroendocrine tumour of duodenum with low mitotic activity and lack of necrosis.
2. Multiple neuroendocrine tumours of mesentery
3. Resected ends of duodenum and jejunum were free of tumour
4. Gall bladder, pancreas and pylorus were free of tumour

The Further histopathology block was sent for Immunohistochemistry and KI 67 to enable exact grading that showed mitotic index less than 1, that was suggestive of a low grade, well differentiated tumour, following which patient was discharged on feeding jejunostomy and oral feeds and followed up after two and four weeks respectively (Figure 5-7).

Discussion

The annual incidence of small intestinal carcinoids is nearly 0.5/100,000, they account for a small proportion of intestinal neoplasms while colorectal adenocarcinomas are 60 times more frequent [6].

Duodenal NETs comprise 1%-3% of all primary duodenal tumors and 2.8% of all carcinoid tumors [6]. The age-adjusted annual incidence of NETs arising from jejunum and ileum is 0.67 per 100000.

Being a rare malignancy, investigation regarding its pathophysiology and information on the classification of these tumors has been limited till recent times. Duodenal NETs are usually diagnosed in the sixth decade with a male predominance [7,8].

There are five types of duodenal NET's

Duodenal gastrinoma [the most common type] duodenal somatostatinoma, non-functioning duodenal NETs, duodenal gangliocytic paraganglioma and poorly differentiated neuroendocrine duodenal carcinomas.

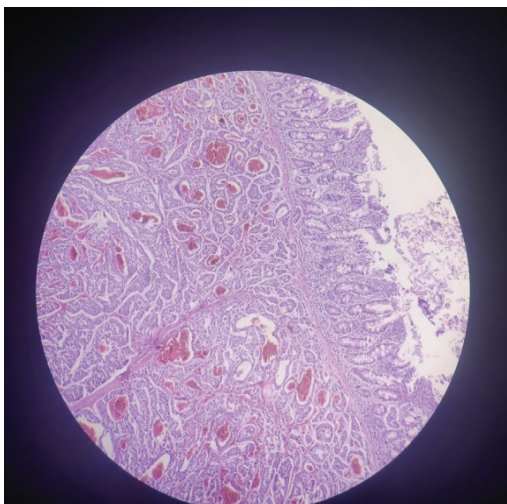


Figure 5: Showing histopathological findings

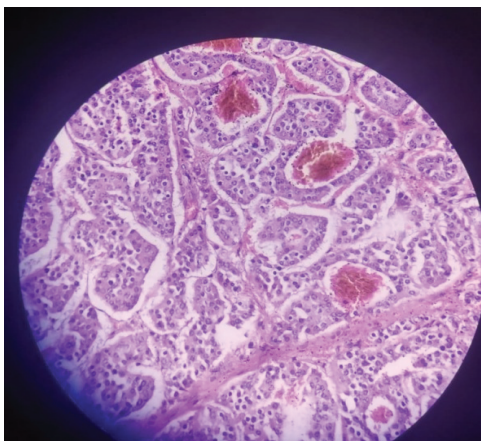


Figure 6: Showing histopathological grading and findings

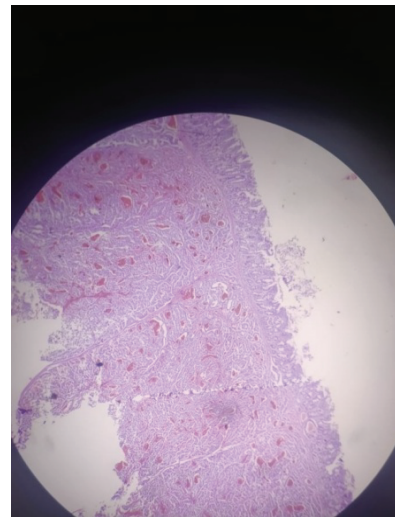


Figure 7: Depicting histopathological findings of neuroendocrine tumor

A Majority of duodenal NET's are small, single lesions, usually limited to the mucosa and submucosa. Regional lymph node metastases may be found in up to 60% of cases, while liver metastasis usually occur in less than 10% [7]. Approximately 90% of duodenal NET's are not associated with clinical syndrome, as majority of diagnosis are made accidentally during a routine work-up or if the patient develops symptoms attributable to the mass itself [7]. Most frequently reported presenting symptoms include pain, jaundice (more frequent in peri-ampullary NET's), nausea, vomiting, diarrhea, obstruction, active bleeding or anemia. In the minority of duodenal NET's that cause a functional syndrome the two main presentations are Zollinger-Ellison syndrome and carcinoid syndrome.

NET's of jejunum and ileum are usually diagnosed in the sixth or seventh decade, but as opposed to duodenal NET's, have no gender preference. Most of them are nonfunctioning tumors, but about 20% of patients show liver metastases and may present with carcinoid syndrome. At diagnosis, lesions are commonly > 2 cm, with invasion of muscularis propria and metastasis to regional lymph nodes. Multiple lesions may be found in up to 40% of cases [9].

USG plays a limited role in the imaging of small bowel NETs. The most common imaging study obtained for the diagnosis of small bowel NETs is a multiphase CT scan.

Gastrointestinal NET's are graded according to the 2010 WHO classification of tumours of the digestive system based on the proliferative index, which is assessed by the percentage of cells that stain positively for Ki-67 and mitotic rate.

The accepted surgical approach for resection of small bowel NET's is an open abdominal operation, to achieve the goals of careful palpation of the entire small bowel and adequate resection of mesenteric lymph nodes while preserving vascular inflow and outflow to the remainder of the intestine.

The need for resecting the primary tumour is to treat or avoid the situations that lead to symptoms, that is, bowel obstruction, bleeding, mesenteric fibrosis, peritoneal dissemination and reducing the risk of further metastasis.

In our case there was a vague clinical presentation of an abdominal lump with only dull aching pain for which a preoperative diagnosis of gastrointestinal stromal tumour was made on cect scan. The tumour proved to be a neuroendocrine tumour on histopathology of the operative specimen which was a rare incidental finding.

Conclusion

Small intestinal NET's are highly prone to metastasize, are fairly

slow growing and associated with relatively favourable survival durations compared to other metastatic cancers.

Regarding prognosis, patients with well-differentiated duodenal NET's have an average global 5-year survival rate of nearly 85% [10]. It is 65% for patients with localized disease and only 36% for those with distant metastasis [11].

The stage of disease at diagnosis highly influence prognosis, with a 10-year survival of 95% for patients with local disease and 10% for those with distant metastases [11].

The prognosis of these NETs is generally unfavourable when compared with other location of tumours of comparable size since they have a higher tendency to grow and spread before the diagnosis is confirmed [12,13].

The diagnosed incidence of SINETs has tripled over the last three decades, likely from frequent use of abdominal radiologic imaging and endoscopic procedures.

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