ORIGINAL RESEARCH PAPER

General Surgery

AN OVERIEW OF FOREGUT CARCINOID – OCCURING AT THE JUNCTION OF SECOND AND THIRD PART OF DUODENUM- A CASE REPORT

KEY WORDS: Duodenal Carcinoid, Foregut Carcinoid, Duodenal Neuroendocrine Tumours, D-nets.

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ABSTRACT

Duodenal carcinoid tumors are uncommon and very rare. Primary duodenal carcinoid lesions account for less than 5% of all gastrointestinal carcinoid tumors. They are sub-classified as Foregut Carcinoids. duodenal carcinoids often present signs of partial duodenal obstruction with symptoms of vomiting, abdominal pain or bleeding in the digestive tract. If it affects the ampulla of Vater, it can cause obstructive icterus or recurrent acute pancreatitis . Symptoms of carcinoid syndrome with the secretion of vasoactive substances are exceptional. We report a case of 57 year old female with vague abdominal pain and malena and was diagnosed as duodenal carcinoid in the third part of duodenum. We present this case to highlight an uncommon and rare foregut carcinoid .

INTRODUCTION:

Karzinoide" (carcinoma-like, lack of malignant potential or particularly benign features). Carcinoids arise from enterochromaffin cells (Kulchitsky cells) . Histologically and biochemically diverse tumors .Neuroendocrine tumors (NETs) of the gastroenteropancreatic (GEP) system are defined as epithelial neoplasms with predominant neuroendocrine differentiation. NETs of the duodenum are considered a distinct entity from tumors of jejunum and ileum . D-NETs are mostly non-functional and often discovered incidentally during a routine upper gastrointestinal endoscopy for other indications . Although primary d-NETs are rare, slow growing neoplasms with indolent clinical behavior, they can be potentially malignant . These tumors tend to spread to the submusosal layer even during the early stages of the disease. D-NETs mostly present as solitary lesions confined to the mucosa and submucosa layer within diameter less than 2 cm. D-NETs are usually multiple in patients with MEN-1 syndrome. Carcinoids of the duodenum present 2% of GEP-NETs and 1 to 3% of all duodenal tumor.

CASE REPORT:

A 57 year old female presented with complaints of vague abdominal pain on and off for the past one year. she also presented with 3 episodes of malena in the past six months. She also had occasional complaints of dizziness , fatigue and fluctuation of blood pressure. She is a known case of systemic hypertension for the past 5 years , history of hyperuricemia and osteoarthritis of both knees. No other comorbidities. On general examination , she was thin built and anaemic. Abdomen was soft with mild tenderness over epigastrium and right hypochondrium. Per rectal examination did not reveal any abnormality. Routine blood investigations were within normal limits with slight decrease in haemoglobin . Upper GI endoscopy **revealed an ulcerated lesion extending beyond D2 and multiple ulcers, Biopsy revealed neuroendocrine tumour.** CE-CT abdomen **revealed a well defined submucosal lesion showing significant enhancement seen at junction of second and third part of duodenum measuring 18*15 mm.** She was evaluated and her hemoglobin levels and blood pressure were corrected as per physician advice and was found to have no other co-existing endocrine abnormalities. The nature of the disease was explained to the patient and her attenders in their understandable language and was taken up for surgery- Laparotomy and proceed – Abdomen opened in layers, all organs inspected, Duodenal loop identified. Kocherisation (mobilization) of duodenum done. Tumour identified – present submucosally over lateral aspect of D2-D3 junction. Tumour removed intoto after duodenotomy . Dudenotomy opening sutured submucosally with 2-0silk, Seromuscular sutures taken along duodenum with 2-0 silk. Omental patch kept and sutured with 2-0 silk. Hemostasis secured, drain placed and transfixed. Abdomen closed in layers. Skin closed in layers. Postoperative period was uneventful. **Tumour sent for HPE revealed as findings consistent with nature of Carcinoid tumour which is positive for Cytokeratin, Synaptophysin, Chromogranin .Ki67 1-2% expression.** Post op Medical Oncologist opinion sought and patient councelled .

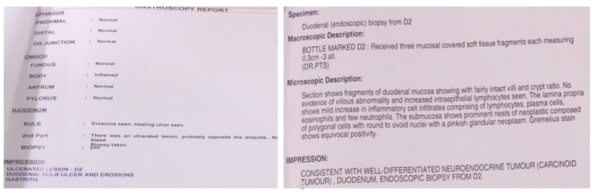


Figure -1 : UGI Report Showing Lesion At D2 And Ugi Biopsy Report Showing Features Of Well Differentiated Duodenal Carcinoid Tumour

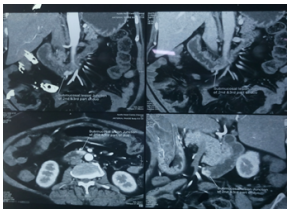


Figure -2: Ce-ct Abdomen Showing A Well Defined Submucosal Lesion Showing Significant Enhancement Seen At Junction Of Second And Third Part Of Duodenum Measuring 18*15 Mm.

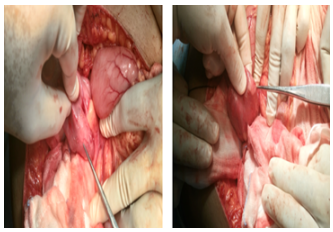


Figure -3: Identification Of Submucosal Tumour At D2-d3 Junction

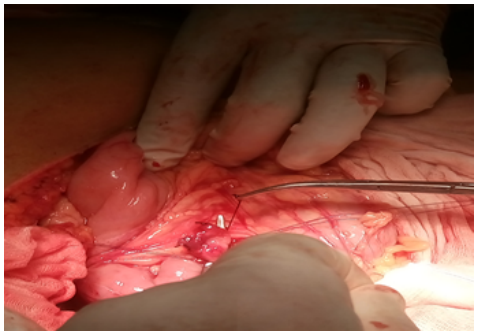


Figure -4 : Duodenotomy And Removal Of Sub Mucosal Tumour At D2-d3 Junction

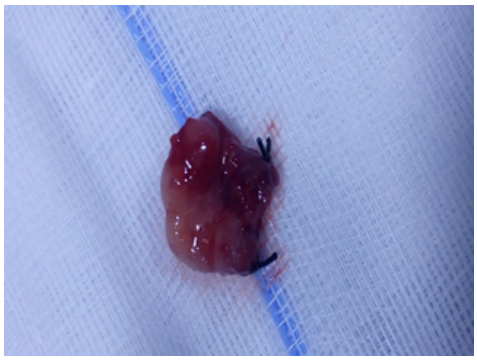


Figure – 5: Removal Of Submucosal Duodenal Tumour.

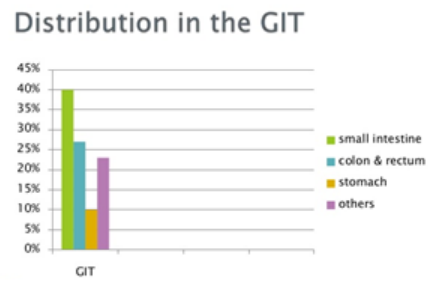
DISCUSSION::

Karzinoide" (carcinoma-like, lack of malignant potential or particularly benign features). Carcinoids arise from enterochromaffin cells (Kulchitsky cells) . Histologically and biochemically diverse tumors. It was first described by Labarsch in 1888. Oberndorfer coined term Carcinoid in 1907. Neuroendocrine tumors (NETs) of the gastroenteropancreatic (GEP) system are defined as epithelial neoplasms with predominant neuroendocrine differentiation Although primary d-NETs are rare, slow growing neoplasms with indolent clinical behavior, they can be potentially malignant . These tumors tend to spread to the submusosal layer even during the early stages of the disease, Neuroendocrine tumors of the duodenum (d-NETs) present 2% of GEP-NETs and 1 to 3% of all duodenal tumors. D-NETs mostly present as solitary lesions confined to the mucosa and submucosa layer within diameter less than 2 cm. D-NETs are usually non-functional, sporadic and well-differentiated slow-growing tumors. Nonfunctional d-NETs are mostly incidentally discovered during an upper gastrointestinal endoscopy. The most common symptoms that lead to diagnostic work-up are abdominal pain (37%), upper gastrointestinal bleeding (21%), anemia (21%) and jaundice (18%)

EPIDEMIOLOGY:

Peak incidence within the sixth to seventh decadeOccur in GIT, Lung, Pancreas, Ovary, Biliary Tract and other soft tissues>50% are found within the GI tract, with the appendix as a very common site of originIncidence is increasing Carcinoid is more common than Small Bowel Adeno Carcinoma

Tumor Site	% of Cases
GI tract	54.5
Small intestine	44.7
Rectum	19.6
Appendix	16.7
Colon	10.6
Stomach	7.2
Lung/Bronchus	30.1
Pancreas	2.3
Gynecologic/Ovarian	1.2
Biliary	1.0
Head and neck	0.4
Other (soft tissue, etc.)	9.7



Characteristics of carcinoids

	Foregut	Midgut	Hindgut
Localization	Stomach, duodenum, pancreas, bronchus	Jejunum, ileum, appendix, ascending colon	Transverse and descending colon, sigmoid, rectum
Histology	Trabecular	Solid mass of cells	Mixed
Silver Staining	Argyrophil	Argentaffin	Variable
Secretory products	5-HT, serotonin, histamine, multiple polypeptides	Serotonin, prostaglandins, polypeptides	None
Metastasis to liver	Stomach 20-25%	Small intestine 35% Asc colon 60% Appendix 2%	Rectum <10% of tumors >2cm
Carcinoid syndrome	Atypical	Classical	Rare

PATHOLOGY:

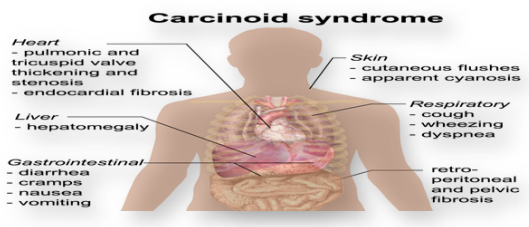
Typical small neoplasm occurring in submucosa. Growth of the tumor is slow; vast majority are <1cm (5%>2cm)It grows outward leaving the mucosa intact .As it reaches the serosa it can cause a desmoplastic reaction and lead to kinking of the bowel. Amplification of HER -2 / neu cellular oncogene expression is seen in ileal carcinoids and obstruction. Metastases occurs in 30% of cases even though the mucosa is intact due to lymphatic spread along the mesentery . Elastic Sclerosis of the mesenteric vessels with Marked fibrosis occurs resulting in angulation and fixation of adjacent bowel loops

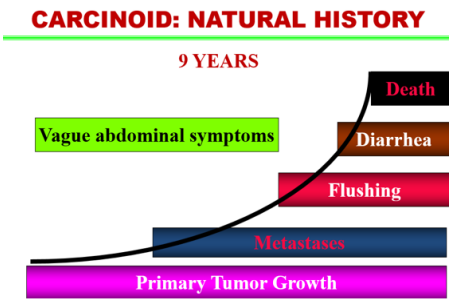
CLINICAL PICTURE:

Usually Asymptomatic It is believed that the average time of onset of symptoms to diagnosis is 9 years. Present with obstructive symptoms. Carcinoid tumours less than 1cm in diameter & confined to the mucosa and submucosa generally remain subclinical for years. Larger than 1 cm – generally malignant & have metastisized to regional lymph nodes and later to the liver and other locations. Patients have generally been complaining for years of intermittent abdominal discomfort, erroneously diagnosed as a functional disorder such as irritable bowel. Intermittent intestinal obstruction due to kinking of the small bowel can occur at a later stage as a result of the desmoplastic reaction in the mesentery . The most common symptoms that lead to diagnostic work-up are abdominal pain (37%), upper gastrointestinal bleeding (21%), anemia (21%) and jaundice (18%) Small tumors are often asymptomatic .30% cause symptoms of obstruction, pain, bleeding, or the carcinoid syndrome or Carcinoid crisis.

Carcinoid symptoms and their putative mediators

Organ	Symptom	Frequency, percent	Putative mediator
Skin	Flushing	85	Kinins, others
	Telangiectasia	25	
	Cyanosis	18	
	Pellagra	7	Excess tryptophan metabolism
Gastrointestinal tract	Diarrhea and cramping	75-85	Serotonin
Heart	Valvular lesions		
	Right heart	40	Unknown
	Left heart	13	
Respiratory tract	Bronchoconstriction	19	Unknown





DIAGNOSIS:

Clinical symptoms

Hormone concentrations

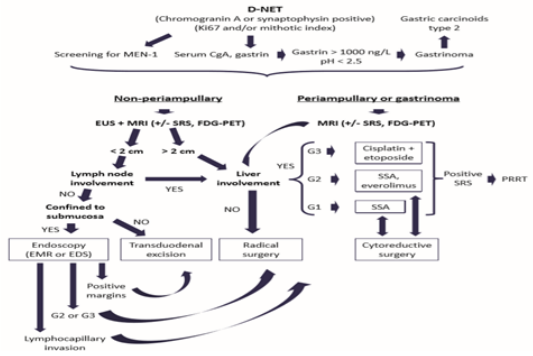
Measurement of urinary or plasma serotonin or its metabolites & 5-HIAA in the urine.
Serum chromogranin A and NSE

Radiology

CT useful in detecting metastases and the extent tumor.
Somatostatin receptor scintigraphy

Histology – gold standard

TREATMENT : ALGORITHM



CONCLUSION:

Duodenal carcinoid tumors are uncommon and very rare. Primary duodenal carcinoid lesions account for less than 5% of all gastrointestinal carcinoid tumors. We present this case to highlight its unique presentation and how it can be diagnosed incidentally and if diagnosed at the earliest, it can be treated and patient can become symptomatically free of carcinoid features. We also present this case to highlight, that there is very little literature about foregut carcinoid occurring at junction of second and third part of duodenum.

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