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Neuroendocrine Tumor of Duodenum : A Rare Case Report

KEYWORDS

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Introduction

Neoplasms of enterochromaffin /neuroendocrine cell origin with neurosecretory capacity that may result in the carcinoid syndrome called as neuroendocrine tumors (NET). Previously NET was called as carcinoid tumors. In 1907, the term carcinoid was introduced by Oberndorfer to describe a tumour of the gastrointestinal tract that was less aggressive than adenocarcinoma and is undergoing a location change within the gastrointestinal tract. Changes in diagnostic modalities used as well as reporting techniques have been observed due to alteration in location of NET.[1] In the United States, estimated incidence ranges from 2.5-5 cases per 100000.[2] About 60% of all cases the gastrointestinal tract is The most common site of primary NETs while 27% incidence was observed in bronchopulmonary tree. Ovaries, testes, liver, biliary tract and pancreas are less frequent sites. The prominent site for incidence of NETs is the small intestine (34%), followed by the rectum (23%), colon (19%), stomach (7.7%), pancreas (7.5%) and appendix (6.6%) observed in the gastrointestinal tract. [3]

CASE REPORT :

We report a rare case of neuroendocrine tumor of duodenum. A 47 years old male came with complain of right sided abdominal pain, anorexia and weight loss. Patient was advised for ultrasonographic evaluation of abdomen and pelvis, large inhomogenous lesion found associated with pancreas and liver. For further investigations CT scanabdomen with biopsy has been performed. CT Scan showed mass posterior to second and third part of duodenum a large 11.2X9.3X6.2 cms sized detected. The lesion was vascular and few perilesional venous collaterals were identified. The mass indents on head and uncinate process of pancreas, posteriorly it indents infrahepatic IVC and posteriorly abutting right kidney. Invasion to adjacent structures was not detected. Four linear tissue cores collected in paraffin block. Immunohistochemistry of the tumor revealed that Chromogranin was strongly positive whereas synaptophysin was weakly positive in tumor cells. Duodenal biopsy with histopathological findings confirmed that mass was well differentiated neuroendocrine tumor.

After thorough pre operative investigations, patient posted for exploratory laparotomy under general anaesthesia and excision of tumor. Tumor was excised totally from duodenum without any resection of bowel. Patient recovered steadily after surgery.

DISCUSSION :

Previously known carcinoid tumor, a duodenal neroendocrine tumor is originated from enterochromaffin cells, and located in gastrointestinal tract. In the United States, only 2.6% duodenal neuroendocrine tumors were diagnosed out of all carcinoid tumors. It became difficult to perform endoscopic treatment for this tumor because of its rare incidence.[4] In the past 35 years, the number of neuroendocrine tumors/carcinomas of the small intestine has increased 300%-500% is reported by the US's Surveillance Epidemiology and End Results (SEER) Register. [5, 6] For the period between 1992 to 2006, mean annual incidence rates per 100 000 population of neuroendocrine tumor of small bowel was 1.00 and 0.70 for men and women respectively reported in the U.S. As per histological assessment, 35%-42% of neoplasms found in the small intestine, most of which occur in the ileum and rarely in the duodenum.[7] 1 to 2 cases per 100,000 people is an estimated prevalence of neuroendocrine tumors, of which gastrointestinal (GI) tract is the most common site.

Approximately, 67.5% is the incidence of GI –NET is around amongst all NET. Limited data is available on epidemiology of GI – NET in India. In a retrospective study, Amarapurkar DN et al reported that a male gender showed preponderance (ratio of 2.5:1) with a mean age of 53.01 ± 15.13 years. As per clinic pathological analysis, out of the 74 tumors, stomach 22 (30.2%) was the commonest site, followed by pancreas 17 (23.3%) and duodenum 14 (18.9%) while 3 (4.1%) patients presented with carcinoid syndrome. Demographic data of our case is comparable with this study.[8]A study performed by Hegde V et al., revealed that an increase in incidence of gastric carcinoids (GC) in India was consistent with the reports from Western countries. Atrophic gastritis is associated with increase in number of GC. It is clear that frequency of occurrence of GC in India is increasing over the time.[9]

Identification of primary tumors and metastatic disease can be assessed by CT and MRI with reliable findings. The process of diagnostic imaging starts with endoscopy (duodenoscopy) and a tissue sample for histological diagnosis. In the case of duodenal NETs, endoscopic ultrasound (EUS) should follow. Endoscopic ultrasound is ideal modality in assessing accurate tumor size and depth of invasion. [1, 10]

Patient was advised to undergo FDG PET/CT for accurate diagnosis. Microscopical examination and histopathological assessment showed positive presence of Chromogranin that validated occurrence of rare neoplasm of the duodenum.On immunohistochemistry, chromagranin is strongly positive in tumour cells and synaptophysin is focally weakly positive which confirmed the diagnosis of well differentiated neuroendocrine tumor of duodenum.

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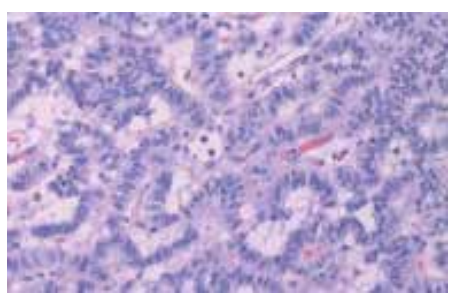
Development of foregut, midgut, and hindgut NETs might involve different genes reported in various studies. Tumor formation may be associated with distinct abnormalities such as mutations, deletions, methylation, and chromosomal losses and gains in proteins like Menin. Menin is a nuclear protein that control transcription regulation, genome stability, and cell division by interacting with cytoplasm with several proteins of dividing cells. Menin mutations are responsible for forgut NETs but still exact molecular etiology of neuroendocrine tumor remain unknown.[5]

CONCLUSION:

Neuroendocrine tumors are frequently detected in gastrointestinal tract. Appendix, ileum and rectum are considered to be common sites for these tumors. Over last few years there has been change in pattern of gastrointestinal neuroendocrine tumors. NET of duodenum is unusual and rare neoplasm. The rapid technological advancements are required for accurate prediction of molecular profile of tumor to identify biomarkers. In the management of early duodenal NETs, predictive reliable biomarkers will have clinical significance. This is rare case report because NET of the duodenum still remaining orphan despite of indentified from last century.

(CT image)



(Histopathology)

(CT image)

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