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Case Report

Neuroendocrine Carcinoma of Unknown Primary - A Rare Presentation

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ABSTRACT

The aim of the article was to present a rare case of metastatic well differentiated neuroendocrine carcinoma of unknown primary in a 20year old female who had presented with complaints of pain abdomen and vomiting. Neuroendocrine tumors (NETs) are rare tumors representing less than 1% of all visceral malignancies. The main primary sites are gastrointestinal tract and lungs. The metastatic sites are Lymph nodes, Liver, Bone, Lungs, Brain and other Organs. Neuroendocrine carcinomas of unknown primary site are uncommon, diverse tumors with variable clinical behavior, predicted by tumor grade or differentiation. In our patient, on ultrasonography solid right mesenteric mass was detected and CT scan showed a vascular mass arising from duodenal wall. Exploratory laparotomy was performed and mesenteric mass was excised. The removed mesenteric mass showed histological and immunohistochemical characteristics of metastatic well differentiated neuroendocrine carcinoma of mesenteric lymph node. Other investigations including repeat CT scan, endoscopy were performed, but no other primary tumor was found. Therefore, we consider our case to represent a neuroendocrine carcinoma of unknown primary.

Key Words: Metastasis, mesenteric lymph node, neuroendocrine carcinoma.

INTRODUCTION

Neuroendocrine tumors are heterogeneous group of neoplasms characterized by embryological, biological and histopathological differences. [(1)] They are rare tumors representing less than 1% of all visceral malignancies and less than 2% of malignant tumors in the gastrointestinal tract. [(2)]

The main primary sites are gastrointestinal tract (62-67%) and the lungs (22-27%). Presentation with metastatic disease accounts for 12-22%. [(3)] Most commonly they metastasize to lymph nodes, liver and less commonly to bone, lungs, brain and other organs. [(4)] Relatively few cases of neuroendocrine carcinoma present with no apparent primary lesion.

CASE PRESENTATION

A 20 year old female presented with pain abdomen since 2 years and vomiting since 6 months. Tenderness in epigastric region was noted on per-abdominal examination. Ultrasonography showed solid right mesenteric mass. Endoscopy revealed

oesophagitis and multiple duodenal ulcers with partial gastric outlet obstruction. Endoscopic biopsy of ulcers was performed. CT scan showed vascular mass arising from duodenal wall in D_2-D_3 curve and thickening of gastric rugal folds (Figure 1). Exploratory laparotomy was done and the excised mesenteric mass was submitted for histopathological examination. On gross examination an encapsulated, light brown mass, firm in consistency, measuring 3x2 cms was noted. Cut surface was solid with light brown to grey white appearance (Figure 2).

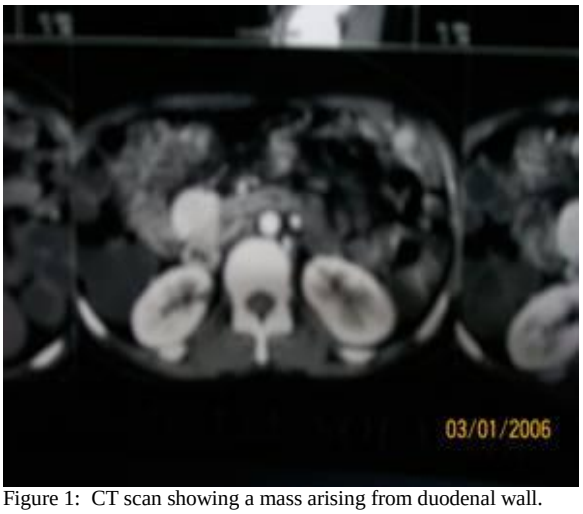

Figure 1: CT scan showing a mass arising from duodenal wall.

Figure 2: Gross photograph - cut surface of the mesenteric mass.

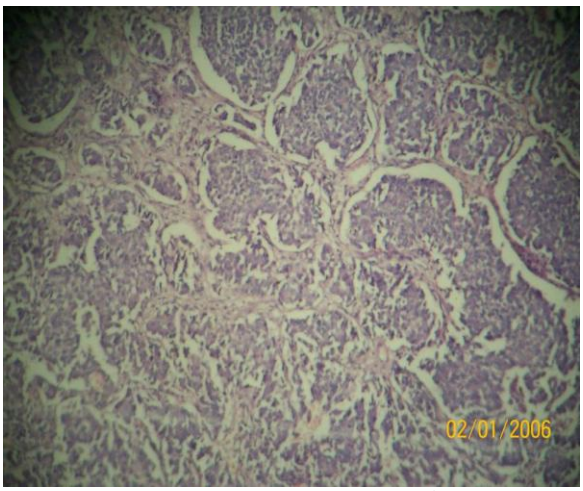

Figure 3: Tumor cells in solid nests separated by fibrovascular Septae (H&E, x 100).

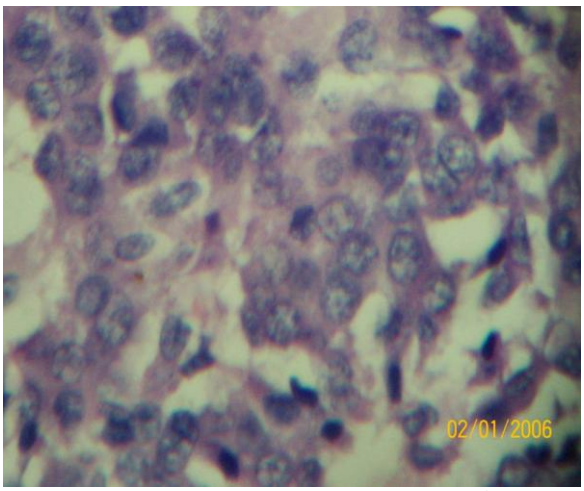

Figure 4: Tumor cells with round to oval nuclei with uniform chromatin and granular eosinophilic cytoplasm (H&E, x 400).

Microscopic examination of the mesenteric mass showed monotonous tumor cells arranged in solid nests and trabecular pattern, separated by thin fibrovascular stroma. Retraction artifacts were noted around the solid nests (Figure 3). Nuclei were round to oval having fine uniformly distributed chromatin with inconspicuous nucleoli. Cytoplasm was granular, eosinophilic and moderate in amount (Figure 4). At places moderate anisonucleosis was noted. 3-6 atypical mitoses/10 HPF were noted. At the periphery, rim of normal lymph node tissue was seen (Figure 5).Endoscopic biopsy of the ulcer edge of gastrojejunal stomal site showed chronic non specific inflammation (Figure 6).

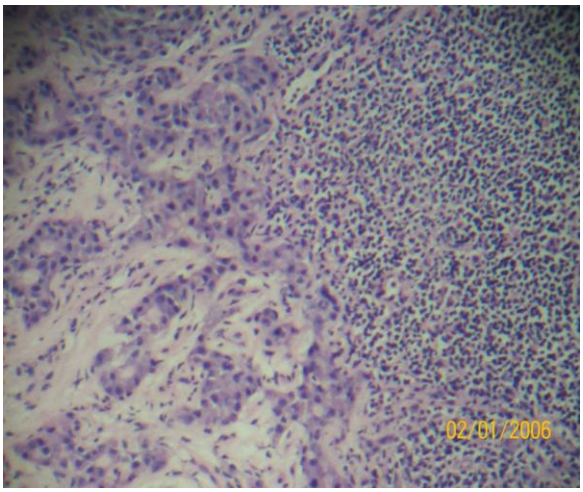

Figure 5: Lymph node and tumor tissue (H&E, x 100).

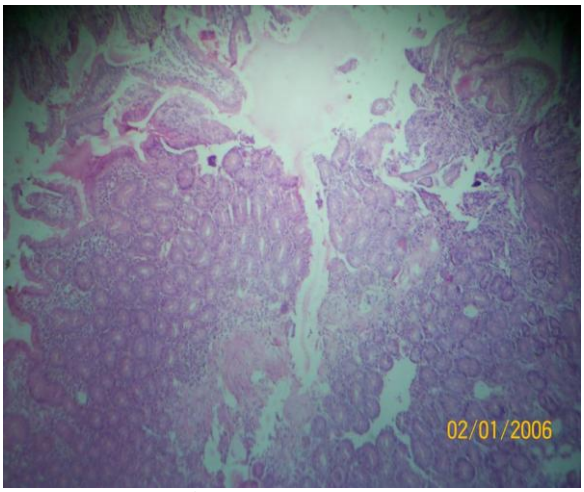

Figure 6: Ulcer edge from gastrojejunal stomal site showing nonspecific inflammation (H&E, x 100).

A differential diagnosis of metastatic carcinoid/ metastatic neuroendocrine tumor/metastatic adenocarcinoma of mesenteric lymph node were given. On immunohistochemistry (IHC) synaptophysin, chromogranin and cytokeratin were positivity while EMA and c-Kit were negative. Based on microscopy and IHC, a diagnosis of well differentiated neuroendocrine carcinoma was made.

DISCUSSION

Well differentiated tumors are the most predominant form of NETs of the digestive tract. They may be either benign or malignant. [(1)] Diagnosis of malignancy in neuroendocrine tumors is suggested by the presence of features such as metastasis, tumor size (tumors>2cm are more aggressive), invasion into adjacent tissue, invasion beyond the submucosa and angioinvasion etc. [(1)]

Neuroendocrine carcinomas of unknown primary site are uncommon and most of them probably arise from an occult/clinically undetectable primary site in one of several locations (bronchus, pancreas, stomach, colon, rectum and several other sites). [(5)] Diagnosis of metastatic well differentiated neuroendocrine carcinoma (NEC) of mesenteric lymph node was made in our case as no primary tumor was detected on various investigations like USG, CT scan and IHC.

Van Boghossian et al [(4)] gave two possible explanations for this. First, the precursor cells in lymph node may become neoplastic leading to primary NET of lymph node. Another explanation is primary might have regressed spontaneously leading to presentation with single site of metastasis. Similar explanation applies to the present case. Further, authors explain that as the precursor cells were not identified within the lymph node, the possibility of metastases was more acceptable.

CONCLUSION

To conclude primary nodal neuroendocrine carcinoma follows a less aggressive course than the metastatic form of nodal involvement. The aggressive nature of this disease necessitates frequent follow-up.

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