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Case Report

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NEUROENDOCRINE CARCINOMA OF OESOPHAGUS: A RARE CASE REPORT DIAGNOSED ON BIOPSY

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ABSTRACT

Neuroendocrine carcinoma (NEC) is a relatively rare disease with a incidence between 0.4% and 2% among all malignancies of the esophagus. We, report a case in which patient presented with dysphagia and diagnosis of oesophageal neuroendocrine carcinoma(NEC) was established based on histomorphological features and immunohistochemical study. On IHC, synaptophysin and chromogranin were strongy positive. The clinical course of oesophageal neuroendocrine carcinoma is very grave, so need for early diagnosis is essential so that treatment could be started at a initial stage of carcinoma.

KEYWORDS: Oesophagus, Neuroendocrine carcinoma, IHC.

INTRODUCTION

Neuroendocrine carcinoma (NEC) is a relatively rare disease with a incidence between 0.4% and 2% among all malignancies of the esophagus.[1] Men are affected more than female, more common in age group 40-60years. It is categorized into two morphological types: small cell type and large cell type. The former is more frequent (approximately 90% of total cases).[2] The prognosis of NEC of the esophagus is poor, so, early detection is important for initiation of appropriate treatment. Herein, we report a case of neuroendocrine carcinoma of oesophagus which was diagnosed on biopsy.

CASE REPORT

61-year-old woman was referred to our hospital with the chief complaints of dysphagia, weight loss and vomiting. Endoscopy revealed a huge type-3 tumor on the abdominal esophagus. Biopsy was performed on the same.

On gross examination hard, ulcerated mass measuring 1x1x.5 cm was found arising from lower end of oesophagus, microscopic examination of the mass revealed sheets and nests of small cells with hyperchromatic nuclei and a minimal to moderate amout of cytoplasm, with prominent crushing artifact.{figure1&2}. Differential diagnosis of poorly differentiated carcinoma, small cell carcinoma, neuroendocrine carcinoma was made.

IHC markers Cytokeratin, CD 20, synaptophysin, chromogranin were applied to the tissue. {Figure3,4,5&6}. As evident from the IHC markers synaptophysin and chromogranin showed strong cytoplasmic positivity, while CK and CD 20 were negative, thus the diagnosis of neuroendocrine carcinoma was confirmed.

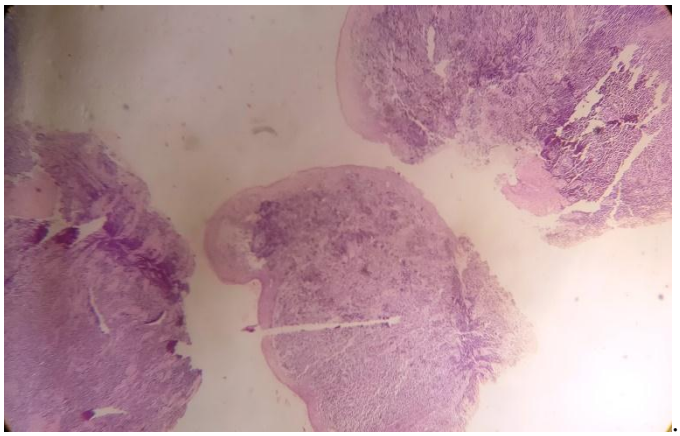

Fig. 1: Monotonous population of tumor cells (H&E,100X).

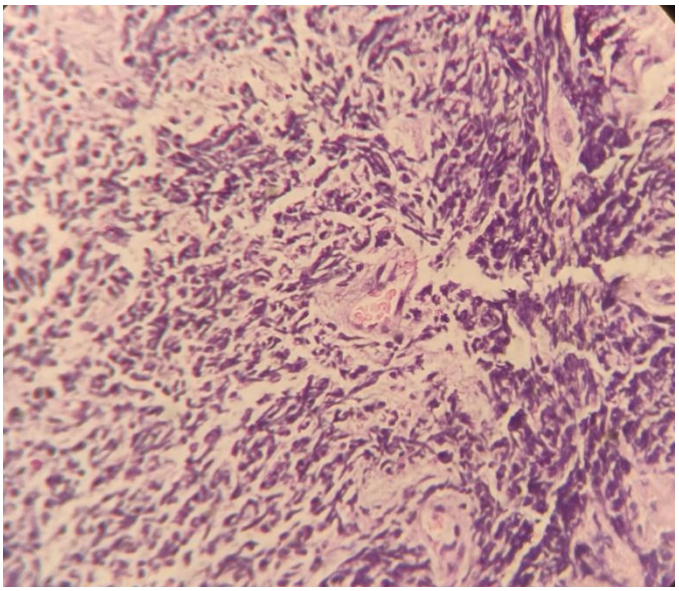

Fig. 2: Sheets of small cells with hyperchromatic nuclei (H&E,400X).

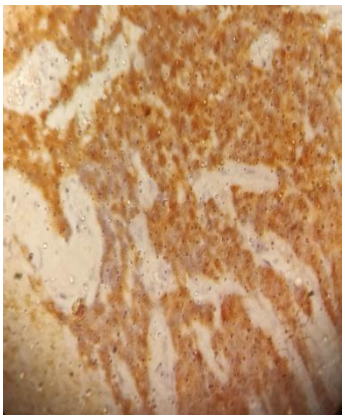

Fig. 3: Synaptophysin positivity(IHC,100X).

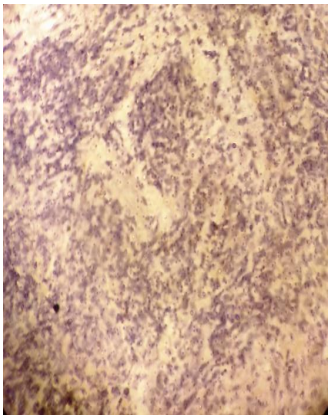

Fig. 5: CD 20 negative(IHC,400X).

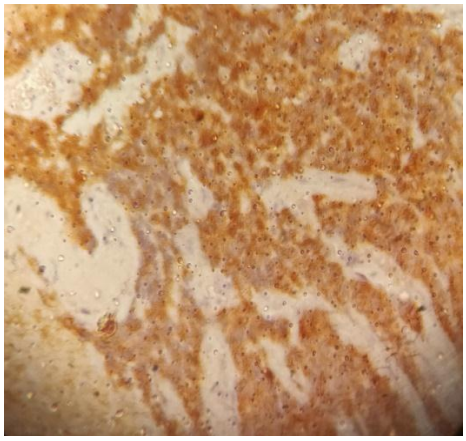

Fig. 4: Chromogranin positivity(IHC,100X).

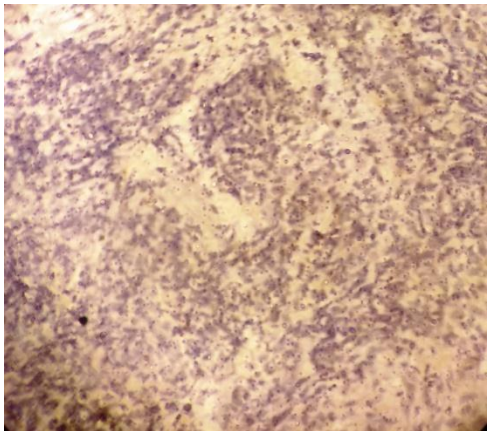

Fig. 6: Cytokeratin negative, IHC, 400X.

DISCUSSION

Neuroendocrine tumors(NETs) are defined as neoplasms that exhibit neuroendocrine phenotypes, such as production of neuropeptides, large dense-core secretory vesicles. According to the World Health Organization (WHO) classification scheme in 2010, NETs can be divided into three main categories: well-differentiated (low-grade), which is benign, medium-differentiated (intermediate-grade) with low-grade malignant behavior, and poorly-differentiated (high grade), which can itself be divided into large cell and small cell neuroendocrine carcinomas (NECs).[3]

WHO Classification for NENs of the GI Tract[4]

Grade	Mitotic Count (per 10 HPFs)	Ki-67 Index, %
G1	<2	<2
G2	2-20	>2-20
G3	>20	>20

NENs have been described in the central nervous system, respiratory tract, the larynx, gastrointestinal (GI) tract, thyroid, skin, breast, and urogenital system. The GI tract and lungs are the most common primary tumor sites.[4]

Neuroendocrine carcinomas are positive for endocrine marker such as choromogranin A, synaptophysin and CD56. Macroscopically, NEC of the gastrointestinal tract presents as submucosal growth, usually covered by normal epithelium with or without ulcerous lesion in the center. Microscopically, tumor cells shows nested and trabecular growth pattern with peripheral palisading and rosette formation. High frequency of venous invasion, lymphatic invasion and perineural invasion are also seen.[3]

Likewise in our case, microscopic examination revealed sheets and nests of small cells with hyperchromatic nuclei and a minimal to moderate amout of cytoplasm with prominent crushing artifact. Diagnosis of neuroendocrine carcinoma was confirmed on synaptophysin and chromogranin.

Esophageal high grade tumors tend to be aggressive. Usually, patients are diagnosed lately, with widespread disease, and with poor prognosis. High grade neoplasms are often regarded as a systemic disease and, just like in lung cancer, chemotherapy is the mainstay of therapy Additional therapy (surgery or radiotherapy) should be considered, but randomized controlled trials still unavailable. For low grade carcinoid, surgical intervention is the treatment of choice.[5]

CONCLUSION

Oesophagus is the rare site to be involved. The clinical course of oesophageal neuroendocrine carcinoma is very grave, so need for early diagnosis is essential so that treatment could be started at a initial stage to improve prognosis of patient.

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