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NEUROENDOCRINE CARCINOMA OF ESOPHAGUS-PRESENTATION AND MANAGEMENT: CASE SERIES

Oncology

Dr.D.J.Kalita	Assistant professor, Department of surgical oncology Dr.B.Borooah Cancer Institute, Guwahati-781008
Dr.Srinivas Bannoth*	Mch Surgical Oncology Dr.B.Borooah Cancer Institute, Guwahati-781008 *Corresponding Author
Dr.Anupam Sarma	Professor, Department of pathology Dr.B.Borooah Cancer Institute, Guwahati-781008
Dr.Niju Pegu	Mch Surgical Oncology Dr.B.Borooah Cancer Institute, Guwahati-781008
Dr.Jitin Yadav	Mch Surgical Oncology Dr.B.Borooah Cancer Institute, Guwahati-781008
Dr.Pritesh Singh	Mch surgical oncology Dr.B.Borooah Cancer Institute, Guwahati-781008
Dr.Dwipen Kalita	M.S .General surgery Dr.B.Borooah Cancer Institute, Guwahati-781008

ABSTRACT

BACKGROUND: Neuroendocrine tumours of esophagus are a relatively rare disease with aggressive biology of tumour. We here present clinicopathological features and management of three cases.

INTRODUCTION: Neuroendocrine tumours of esophagus are rare tumours with incidence of 0.04-1.4 % of gastro intestinal neuroendocrine tumours(NETs) **(1,2)**.The WHO classification system divides neuroendocrine neoplasms (NENs) into low to intermediate grade(grade1-2) NETs and high grade (grade 3) neuroendocrine carcinomas(NECs ,large or small cell type) **(3,4)**. Classified into two categories based on clinical symptoms and levels of hormone secretion into functional and nonfunctional **(5)**.NETs of esophagus are found more commonly in men in the sixth decade of life **(6)**.

KEYWORDS

Neuroendocrine tumours, esophagus,management

CASE PRESENTATION:

CASE CAPSULE 1. 47 year female with ECOG-1 status (Eastern cooperative oncology group) came with chief complaint of dysphagia to solids for 15 days with generalized weakness. On examination there was no pallor, icterus, cyanosis or lymphadenopathy. Blood work up was done which was with in normal limits.Upper gastrointestinal endoscopy (UGIE) done which was suggestive of growth from 30 to 34 cm, from upper incisors. Contrast enhanced computerized tomography(CECT) of thorax and abdomen done which was suggestive of Carcinoma of mid and lower thoracic esophagus with disseminated hepatic metastasis ,enlarged mediastinal nodes and conglomerate metastatic nodal mass in gastrohepatic space.Biopsy and Immunohistochemistry (IHC) was suggestive of neuroendocrine tumour of esophagus (Chromogranin A, synaptophysin and cytokeratin positivity).Multidisciplinary tumour board(MDTB) done and was planned for palliative chemotherapy.Six cycles of chemotherapy given etoposide and cisplatin. Patient had subjective improvement in dysphagia and was kept on best supportive care.

CASE CAPSULE 2: 63 year male ECOG-1 came with dysphagia for one month which was progressive .On examination there was no pallor, icterus or lymphadenopathy. Blood work up done which were with in normal limits.UGIE suggestive of growth from 30 to 35 cm from incissors.CECT thorax and abdomen done which was suggestive of mid thoracic esophageal growth commencing from 1 cm below carina for a length of 7 centimeters, abutting left main bronchus, main pulmonary artery and left atrium with aortic encasement of < 90 degrees. Biopsy and IHC was suggestive of neuroendocrine tumour with positivity of chromogranin A, synaptophysin and cytokeratin positivity).MDTB was done and was planned for chemotherapy followed by radiotherapy plus chemotherapy.Three cycles of etoposide plus cisplatin given followed by External beam radiotherapy of 60 gray in 30 fractions with 5 cycles of concurrent cisplatin. Patient had subject improvement of dysphagia but after 2 months presented with generalized body ache and weaknes .Patient was referred to palliative medicine for best supportive care.

CASE CAPSULE 3: 51 year male came with dysphagia for 10 days and generalized weakness, on examination his general condition was poor after stabilization patient was evaluated .UGIE suggestive growth in around 20 cm from incisors. CECT thorax and abdomen was suggestive of circumferential wall thickening of around 8 cm in upper to m id-part of thoracic esophagus with mediastinal and lung invasion and mediastinal lymphadenopathy. Biopsy was suggestive of poorly differentiated squamous cell carcinoma with neuro-endocrine differentiation.With cytokeratin, chromogranin A, and synaptophysin positivity. After MDTB patient was planned for palliative radiotherapy.

Figure 1: CECT image showing esophageal thickening

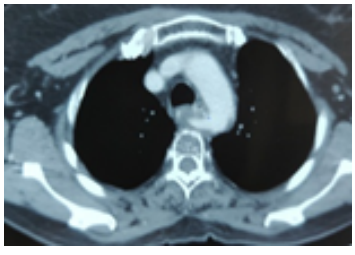
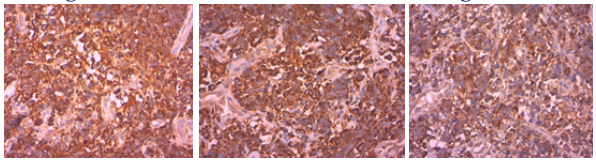


Figure 2: Synatophysain staining

Figure 3: cytokeratin staining

Figure 4 Chromogrannin staining



DISCUSSION:

Esophageal neuroendocrine tumours are rare type of esophageal tumours which are usually positive for chromogranin A , synaptophysin, and CD56.Esophageal neuroendocrine tumours are rare comprising only 1.4% of all gastroenteropancreatic neuroendocrine tumours**(2).**The reported incidence and prevalence of NETs have increased due to improved diagnostic techniques**(7).**

The demographic data of patients with esophageal NEC revealed a mean age of 58.4 years and a male predominance similar to that of esophageal squamous cell carcinoma **(9).**

Most commonly reported symptoms of esophageal NETs are

gastrointestinal symptoms such as dysphagia and abdominal discomfort **(2)**, our patients also presented with dysphagia and generalized weakness.

Limited number of cases has been reported in literature. NETs of esophagus appear to confer worst prognosis as seen in our cases than other esophageal cancers, probably due to its poor differentiation and lymphovascular invasion.

Lee et al proposed treatment algorithm for esophageal NETs in which patients with widespread metastasis receive palliative chemotherapy mainly with cisplatin**(2).**A tumour size > 2cm and American Joint committee on cancer TNM(Tumour extent, lymphnode spread and metastasis) advanced stage have been reported to be poor prognostic factor for overall survival of patients with esophageal NETs**(2,8).**

Therapeutic strategy of NEC of esophagus has not been well defined due to small number of cases reported in literature so far **(10).**NEC can be categorized into two groups one is limited disease(LD) and other extended disease(ED).For ED chemotherapy is the predominant treatment strategy and sometimes radiotherapy.For LD therapeutic strategies are more complicated.Surgery plays the main role hower multimodality treatment such as surgery followed by adjuvant chemoradiotherapy or neoadjuvant chemotherapy followed by surgery are commonly recommended**(10).**

Conclusion: Esophageal neuro-endocrine tumours are rare neoplasms with aggressive behavior whose treatment strategy is yet to be well defined.Multi-disciplinary approach is needed for better outcomes of patients. Surgery plays main role in limited disease with neo-adjuvant and adjuvant treatment and palliative chemotherapy and radiotherapy in case of extensive disease.

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