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Advanced esophageal neuroendocrine carcinoma

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Dear Editor,

Neuroendocrine tumors of the esophagus represent 0.04-0.1 to 1.0 % of all neuroendocrine tumors (NETs) (1-3), and mainly affect males in their sixties, involve the middle and lower esophageal thirds, and manifest primarily with dysphagia. At diagnosis they are usually advanced, highly aggressive small-cell (90 %) or large-cell (10 %) neuroendocrine carcinomas (NECs) (2,3).

Case report:

A 65-year-old patient with progressive dysphagia and toxic syndrome underwent upper GI endoscopy, which revealed a stenosing lesion in the middle and lower thirds of the esophagus that the endoscope could pass. Biopsy showed a high-grade NET with Ki67 at 70%. The lesion was staged as G3 NEC. The study was completed with abdominal ultrasound and CT, their images showing multiple liver metastases that grew rapidly. No carcinoid syndrome (CS) was observed.

He was managed with chemotherapy and died from liver failure within 1 year after diagnosis.

Discussion

Recently, four NEC cases (3-5) were reported in your Journal, which are summarized and compared in Table 1.

As may be seen in this succint review, esophageal NEC involved males over 60 years of age who had predominantly dysphagia. Most cases received chemotherapy because of advanced disease.

Treatment has not been standardized; for a lesion smaller than 1 cm, unusual, occasionally associated with Barrett's esophagus (3), endoscopic resection will be curative; however, all other lesions require recourse to neoadjuvant chemotherapy and surgery in order to prolong life expectancy. Only one case is reported in the literature as being disease-free after 5 years.

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Table 1.

NEC Age	Sex	Presentation	Lesion	Site	NEC	Ki67	Imaging	Therapy	Survival
(2) 65	Male	Dysphagia	Ulcerated	Lower third	Small-cell	53 %	CT	Chemotherapy	Death
Case 65	Male	Dysphagia	Stenosing	Mid-Lower	Small-cell	70 %	CT/US	Chemotherapy	Death
(3) 65	Male	Pain	Ulcerated	Cardia	Large-cell	70 %	EUS/CT	Surgery	6 months
(4) 61	Female	Dysphagia	Ulcerated	Mid third	Small-cell	80 %	CT	?	?
(5) 68	Male	Dysphagia	Polypoid	Mid third	Large-cell	90 %	EUS/CT	Chemo + Surgery	
(5) 55	Female	Dysphagia	Stenosing	Mid third	Small-cell	95 %	CT	Palliative	Death