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Case Report

Esophageal neuroendocrine carcinoma: A clinical perspective on a rare and challenging disease ☆,☆☆

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ABSTRACT

Neuroendocrine carcinomas (NECs) of the esophagus are exceptionally rare, comprising only 0.04% of all neuroendocrine tumors. These malignancies can remain asymptomatic or manifest through digestive symptoms such as dysphagia and abdominal discomfort. Due to their rarity, limited studies exist, resulting in insufficient knowledge about their prognosis and treatment, with conflicting data in the literature.

This report presents the case of a 62-year-old male chronic smoker with chronic dysphagia. Diagnostic evaluations, including gastroscopy and CT, revealed a circumferential lesion in the lower esophagus. Histopathological analysis confirmed a diagnosis of poorly differentiated large-cell neuroendocrine carcinoma.

Esophageal NECs are aggressive and poorly understood tumors. Tumor size, lymph node involvement, and metastatic status are critical factors influencing treatment decisions. However, there are no established guidelines for their management. Extensive research is urgently needed to develop standardized treatment approaches and improve the prognosis of patients with high-grade esophageal NECs.

Abbreviations: CT, Computed tomography; NEC, Neuroendocrine carcinoma.

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Introduction

Neuroendocrine carcinomas (NEC) of the esophagus are extremely rare, as they represent 0.4% to 2% of all esophagus tumors [1], and 0.04% of all neuroendocrine tumors [2].

While the most commonly reported symptoms are dysphagia or abdominal discomfort, some tumors might be discovered incidentally during endoscopy.

With very few studies found in the literature, the knowledge surrounding NEC is reduced with the lack of standardized guidelines for its treatment and conflicting data regarding its prognosis [3].

Due to improved diagnostic techniques, NEC have become an important diagnostic differential to consider, with incidence rates gradually increasing since 1970s [4].

We report the case of a 62-year-old man, admitted for chronic dysphagia, to which the diagnosis of a NEC of the esophagus was retained.

Case presentation

A 62-year-old man, with history of chronic smoking for over 40 years, was admitted in our ward for chronic dysphagia.

He had a prolonged history of swallowing difficulties with multiple episodes of vomiting accompanied with a weight loss of 20 kgs in the last 6 months.

The patient also endorsed having early satiety, a reduced appetite and food regurgitation.

No family or personal history of neoplasm was found.

His initial vital signs were normal. The physical examination showed a cachectic body habitus with a nondistended abdomen. No hepatomegaly or large palpable gallbladder was palpable.

Other than a 3.3 mmol/l hypokalemia and a 132 mmol/l hyponatremia, routine blood and urine tests were normal.

The patient proceeded to an upper endoscopy, which revealed a hemi-circumferential ulcer-budding process in the lower part of the esophagus.

The biopsy revealed synaptophysin and CD56 immunostaining positive tumor (with a negative CK7 and P40) indicating poorly differentiated large cell neuroendocrine carcinoma (Figs. 1 and 2).

A CT, for extension purposes was performed, revealing a 35 mm stenosing and circumferential process of the lower part of the esophagus, spanning at least 110 mm in craniocaudal dimensions, without any secondary location (Figs. 3 and 4).

After a multidisciplinary concertation meeting, the resection of the mass was recommended.

Unfortunately, right before programming his surgery, the patient died at home with his family.

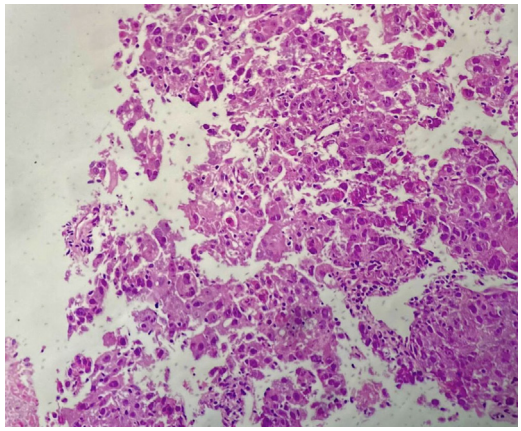

Fig. 1 – Hematoxylin & Eosin stain: large neoplastic cells with distinct nucleoli under 25x magnification.

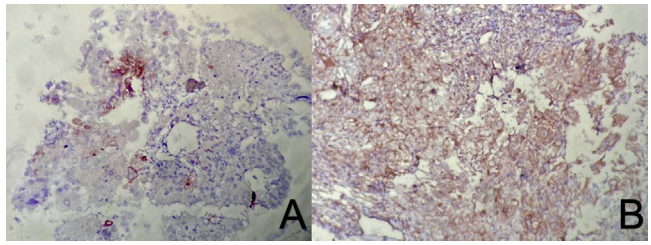

Fig. 2 – Esophageal biopsy showing focal positivity for chromogranin (A) and diffuse positivity for synaptophysin (B) (x25).

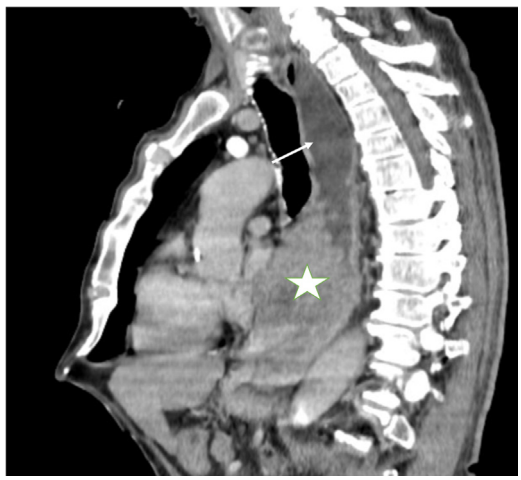

Fig. 3 – CT scan (sagittal plane) showing a lower esophagus mass (white star) with dilation of proximal esophagus above (white arrow).

Discussion

Neuroendocrine neoplasms of the esophagus are quite rare, representing 0.4% to 2% of all esophagus tumors [1]. They are generally classified by their grade; G1 and G2 for low and intermediate-grade neoplasm, and NEC (neuroendocrine carcinoma) for high-grade neoplasms. This last type, characterized by large cell histology, which was the case with our patient, has only been confirmed in less than a dozen patients

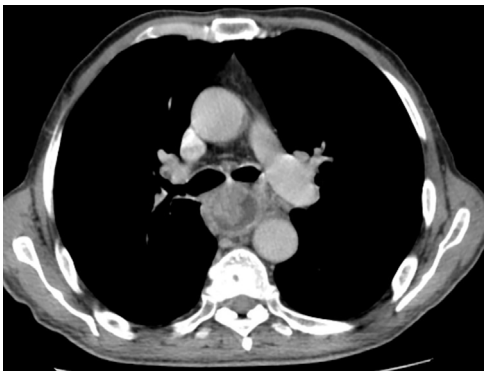

Fig. 4 – Thoracic enhanced CT scan in axial views, revealing a large mass in the distal esophagus, in contact with the left atrium (anteriorly), pulmonary veins, descending aorta (laterally), carina (superiorly), and left main bronchus, without evidence of invasion (white arrows, A and B).

in the literature thus far [5], with a male predominance and a median age of 67 years old.

Due to a higher presence of Merkel and endocrine cells, this tumor usually originate in the mid and lower esophagus [6].

No specific risk factors to esophageal NEC have been defined in the literature, but smoking and alcohol consumption are accepted as key factors, similar to esophageal squamous cell carcinoma [3].

Its anatomical site plays a vital role in its symptomatology. While most of the patients remain asymptomatic, some might experience gastrointestinal symptoms (dysphagia, abdominal discomfort, diarrhea…), respiratory and endocrine symptoms, or classical systemic signs (like asthenia and weight loss) [7]. In some cases, the diagnosis may be made during an endoscopic examination [8]. This initial upper gastrointestinal endoscopy will be used to assess the intra-esophageal expansion and help us provide histological confirmation of the diagnosis.

Due to its rarity, there is currently no consensus about the optimal treatment regimen of NEC. But Lee et al., in the biggest cohort study to date [3], proposed a treatment algorithm based on tumor size and lymph node metastasis. Following this process, our patient, who had a tumor size of 11 cm (>1 cm) without regional lymph nodes metastasis, would fall under surgical resection ± adjuvant chemotherapy. Unfortunately, he died prior to his hospitalization.

Generally, the prognosis for patients with pure esophageal NECs is significantly worse than for those with mixed NECs [9].

In conclusion, NECs of the esophagus are extremely rare, aggressive and poorly understood.

While tumor size, lymph node presence and metastatic disease are important factors in the selection of appropriate treatment, no official management guidelines have been established yet, although general recommendations suggest surgery with adjuvant chemoradiotherapy as the treatment of choice.

Further extensive research is still mandatory to standardize the optimal treatment modalities for high-grade large cell esophageal NEC, so we can improve our patients prognosis.

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Patient consent

Written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article.

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