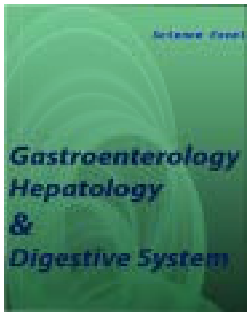
Gastroenterology Hepatology and Digestive System

Case Report



Esophageal Neuroendocrine Tumor: Case Report

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- Received Date: 08 Aug 2022
- Accepted Date: 16 Aug 2022
- Publication Date: 23 Aug 2022

Keywords: neuroendocrine tumors, neuroendocrine carcinomas, esophageal tumors

Abstract

Introduction: Neuroendocrine tumors, also called neuroendocrine carcinomas, are those that form in any region of the neuroendocrine system. Although they are more common in the gastrointestinal tract (67.5%), their appearance in the esophagus is rare. Another important characteristic is its slow growth and low degree of aggressiveness.

Case report: A46-year-old male patient, seeking care for progressive phagia for solidspathstenoses digestive endoscopy that showed the diagnosis of distal esophagus whose anatomoological and immunohistochemical was of a neuroendocrine esophageal tumor. Subjected to staging, there were numerous liver metastases and lymph node enlargement of the celiac trunk, and palliative treatment was chosen. The patient had an unfavorable evolution, progressing to jaundice, ascites, worsening of general condition, progressed to death in about 3 months after diagnosis.

Discussion: The incidence and prevalence of neuroendocrine esophageal tumor has been increasing since 1970 due to the modernization of diagnostic techniques. Despite the increase in diagnostic capacity, this pathology remains rare, corresponding to 0.15% - 2.80% of esophageal carcinomas. The median survival time for patients with this neoplasm is 22.4 months, having a totally different evolution from other neuroendocrine tumors of the digestive tract..

Introduction

Neuroendocrine tumors (NET), also called neuroendocrine carcinomas, are those that form in any region of the neuroendocrine system. When some of the cells in this system undergo a mutation - which can be by chance or hereditary - they start to multiply in a disorderly way, leading to the growth of an abnormal mass of tissue: the neuroendocrine tumor. This can be classified into: well-differentiated neuroendocrine cancers, poorly differentiated cancers and moderately differentiated cancers [1].

Although these tumors can appear in any region of the neuroendocrine system, they are most common in the gastrointestinal tract (67.5%), lungs (25.3%), and pancreas (5.9%).

Other characteristics are its rarity and its slow growth, extending over years. Thus, efforts by researchers to complete meaningful studies to define recommended approaches for all situations are significantly constrained. Since the early 1980s, interest in the study of neuroendocrine tumors has emerged in different centers, with experts coming together to carry out an increasing number of studies, mainly in the treatment of advanced diseases and metastatic stages to overcome the shortage of cases in most practices. clinics [2].

As for the diagnosis of these tumors, when performed at an early stage, it is usually treated surgically, with a good chance of cure. However, the initial cases are asymptomatic, leading to the diagnosis at an advanced stage in the vast majority of patients and, still, present in metastasis [3]

In the case of esophageal NETs, positivity for chromogranin A (or parathyroid secretory protein 1), synaphophysin and CD56 in immunohistochemical staining is common [4].

The aim of this article is to present the case of a patient with a neuroendocrine tumor of the esophagus, a rare pathology that causes dysphagia and/or abdominal discomfort.

Methods

The information contained in this case report was obtained by reviewing medical records, photographing the diagnostic methods to which the patient was submitted and reviewing the literature.

Citation: Mocellin L, Agostinho EP, da Silveira VB, Goncalves MO, Neto JAD. Esophageal Neuroendocrine Tumor: Case Report. Gastroenterol Hepatol Dig Sys. 2022;1(1):1-3.

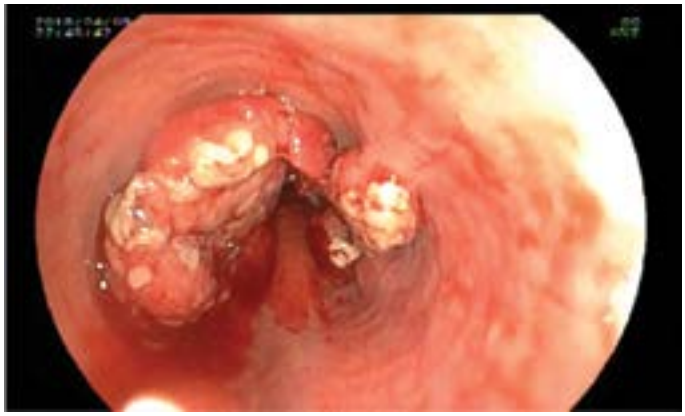

Figure 1. *Endoscopy photograph showing tumor*

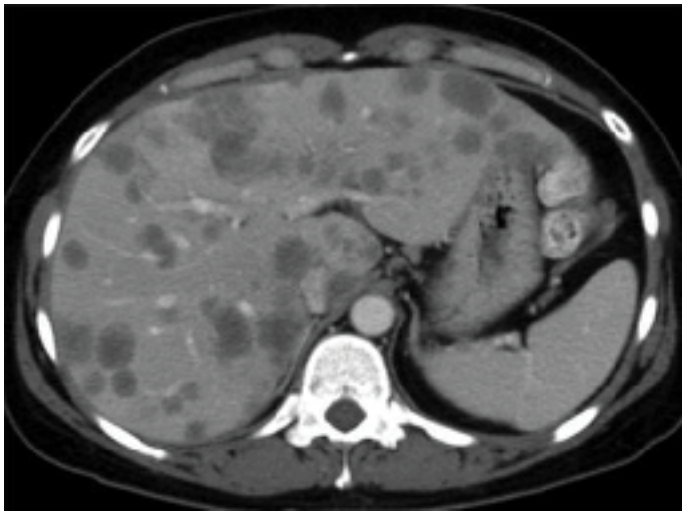

Figure 2. *Tomography showing metastases*

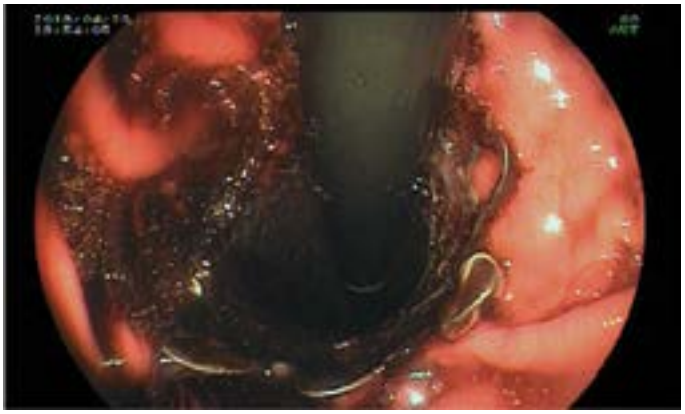

Figure 3. *Endoscopy photograph showing prosthesis in rear view*

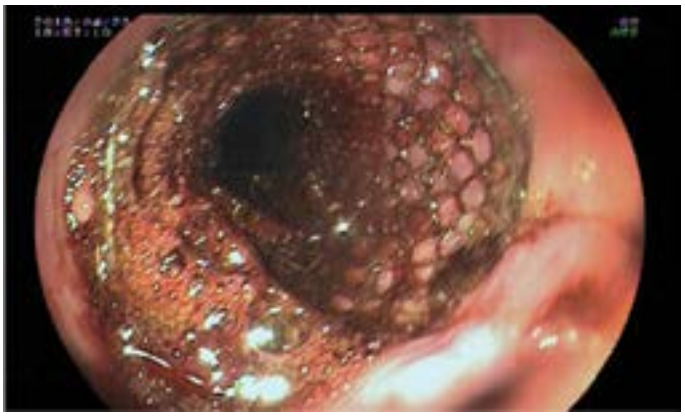

Figure 4. *Endoscopy photograph showing prosthesis in frontal view*

Case report

A 46-year-old male patient, with no previous comorbidities, elongated, sought care from the Digestive System Surgery team due to progressive dysphagia for about 20 days. Refers to dysphagia for solids and tolerates well pasty. Thus, he underwent upper digestive endoscopy (Figure 1).

He underwent staging, which showed numerous metastatic liver lesions and lymph node enlargement in the celiac trunk (Figure 2).

The case was discussed jointly by the Digestive System Surgery, Oncological Surgery and Clinical Oncology team. We opted for the passage of a partially coated esophageal prosthesis associated with palliative chemotherapy. There were no difficulties in introducing the device through the Upper Esophageal Sphincter. Normal mucosa up to 35 cm from the Upper Dental Arcade (ADS), where there is an infiltrative, friable, bleeding lesion, occupying more than 75% of the organ's lumen, which makes it difficult for the device to pass and extends virtually to the esophagus-gastric transition, which located 44 cm from the ADS, already biopsied in another endoscopy. Then, the prosthesis is passed with retrovision (Figure 3) and control endoscopy, which shows a well-positioned and well-expanded prosthesis (Figure 4).

The pathological examination resulted in poorly differentiated malignant neoplasm. Thus, an immunohistochemical study was necessary, which was positive for chromogranin A, synaptophysin and CD56.

The patient had an unfavorable evolution, progressing to jaundice, ascites, worsening of the general condition, and he died about 3 months after diagnosis.

Discussion

The incidence and prevalence of esophageal neuroendocrine tumors have been increasing since 1970 due to the modernization of diagnostic techniques [4]. Despite the increase in diagnostic capacity, this pathology remains rare, corresponding to 0.15%-2.80% of esophageal carcinomas.

The median survival time of patients with this neoplasm is 22.4 months, with a completely different evolution from other neuroendocrine tumors of the digestive tract [5].

Its macroscopic presentation is marked by submucosal growth, usually covered by normal epithelium with or without an ulcerated lesion. at the center [6].

In the microscopic presentation, NETs have great heterogeneity of their cellular components and of the peptidergic products they secrete. Therefore, in 2010, the

World Health Organization (WHO) proposed an update to the classification of NETs that takes into account the degree of differentiation of cells and the proliferative capacity of tumors, assessed by the number of mitoses and the cell proliferation index [7]. Thus, in the microscopy of this case, the sections showed an ulcerated neoplasm constituted by the proliferation of cells containing voluminous, pleomorphic nuclei, with dense chromatin and acidophilic cytoplasm, arranged in blocks of varying sizes, branched and anastomosed, containing several atypical mitotic figures.

For the diagnosis of NET, endoscopy with biopsy, computed tomography or magnetic resonance imaging should be included. In addition, positron emission tomography with fluorodeoxyglucose is indicated for detection of extra-abdominal metastases. As recommended for other types of cancer, complete resections with lymph node dissection can be performed in localized NETs [8].

The reported case and the analyzed publications help the understanding of neuroendocrine tumors, especially those of the gastrointestinal tract. The rarity of the pathology

- in the esophageal location - and its rapid progression, contrary to the literature, justify this report.

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