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(CASE REPORT)

A rare entity: Neuroendocrine tumor of the esophagus about one case

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Abstract

Neuroendocrine carcinoma of the esophagus is a rare, highly aggressive malignancy characterized by rapid progression. The clinical picture is not very specific, ranging from simple abdominal discomfort to dysphagia with altered general condition.

Here, we present the case of a 65-year-old woman with a history of chronic gastroesophageal reflux disease, who presented with dysphagia, exploration of which revealed neuroendocrine carcinoma of the esophagus.

Due to its rarity, the treatment strategy is not yet standardized. Surgery combined with adjuvant therapy appears to be the best option for treating limited-stage esophageal neuroendocrine carcinoma.

Keywords: Neuroendocrine Carcinoma of Esophagus; Dysphagia; Gastroesophageal Reflux Disease; Surgery

1. Introduction

Neuroendocrine tumors can develop throughout the digestive tract, particularly in the small intestine (45%), rectum (20%), appendix (16%), colon (11%) and stomach (7%) [1]. However, esophageal localization remains exceptional.

Given their rarity, little is known about their clinical characteristics, diagnostic methods and available therapeutic strategies. Nevertheless, they are characterized by aggressive behavior, early dissemination and poor prognosis in the absence of treatment [2].

In this report, we describe a case of neuroendocrine carcinoma of the esophagus in a patient who presented with dysphagia.

2. Case report

We report the case of a 65-year-old woman who has suffered from gastroesophageal reflux disease (GERD)for 8 years complicated by peptic stenosis, for which she underwent a single session of hydrostatic balloon dilatation 6 years ago, and Barrett's esophagus for which she was taking long-term proton pump inhibitors (PPIs).

The patient presented to the hospital with progressive dysphagia evolving for 2 months, associated with marked weight loss. The review of symptoms was negative for nausea, vomiting, epigastric pain, and stool character changes.

Clinically, the patient was stable, with a respiratory rate of 20 breaths per minute, a blood pressure of 132/82 mmHg, a heart rate of 88 beats per minute, a weight of 50 kg and a body mass index of 17 kg/m2. The physical

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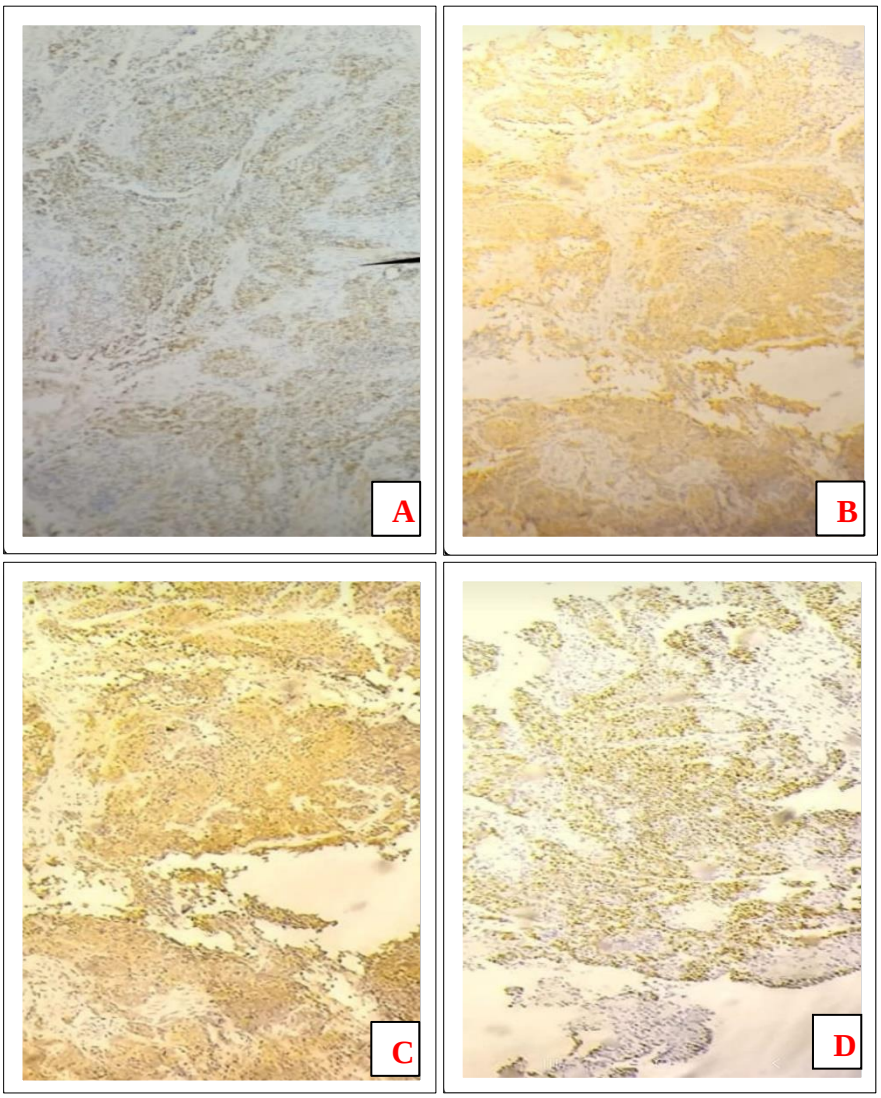
examination was unremarkable. Biological workup was unremarkable, in particular renal function, natremia and kalemia.

An oesogastroduodenal fibroscopy was performed, showing a long Barrett's oesophagus classified as C9M9 according to the prague classification, an irregular stenosis suspected of malignancy extending over 3cm presumed to be 35cm from the dental arches. Hydrostatic balloon dilatation of the stenosis was performed with multiple biopsies.

Biopsies of the stenosis were consistent with esophageal neuroendocrine carcinoma. Tumor cells showed diffuse positivity for CK7 and low expression of synaptophysin. Chromogranin was also expressed by some cells, and complementation by CD56 confirmed widespread positivity of tumor cells. Also, Ki 67 is expressed by almost 60% of cells in the most proliferative sectors



Histological appearance of a poorly differentiated neuroendocrine carcinoma with diffuse positivity of the tumour cells for CK7 (A), expression of synaptophysin (B), positivity of CD 56 (C) and a Ki-67 index expressed in almost 60% of the cells (D)

Figure 1 Immunohistochemistry of neuroendocrine tumor of the esophagus

Thoraco-abdomino-pelvic CT revealed suspicious digestive parietal thickening of the esogastric junction, associated with locoregional lymph nodes and multiple liver metastases.

After in-depth discussion in a multidisciplinary consultation meeting, and given the tumor's locoregional extension and multiple liver metastases, the therapeutic decision was chemotherapy with palliative care. The patient has just had her first chemotherapy session.

3. Discussion

Neuroendocrine carcinoma (NEC) of the oesophagus is very rare disease with a reported incidence between 0.4% and 2% among all malignancies of the esophagus [3]. The World Health Organization (WHO) definition for NEC includes positive endocrine marker such as choromogranin A, synaptophysin and CD56.

Possible risk factors for esophageal NEC have not been extensively studied, but most studies indicate that men are at greater risk of this disease [4].

The clinical picture of esophageal neuroendocrine tumors is similar to that of the other most common histological forms, but is generally more aggressive. Dysphagia is the most frequent clinical symptom, followed by weight loss and/or loss of appetite. Retrosternal and epigastric pain, odynophagia, dysphonia, dyspnea and digestive bleeding (hematemesis and melena) have also been described, although less frequently [2].

Esophageal NEC are usually discovered by chance during endoscopic examination, and develop in the lower third of the esophagus. They usually present as a single lesion, but sometimes as an ulcerated or fungating mass, and deeply infiltrate the lining of the esophagus. Histological diagnosis is established by esophago-gastro-duodenoscopy (EGD) and biopsy with immunohistochemical and histochemical staining for common neuroendocrine markers, including neuron-specific enolase (NSE), synaptophysin, chromogranin A (CgA), CK (cytokeratin), and CD56 (lymphocyte antigen 56) [2].

Endoscopic ultrasound remains the most useful means of assessing the extent of esophageal wall invasion and lymph node malignancy [2]. Imaging also plays an essential role, particularly in assessing distant tumor extension.

Given the low incidence of esophageal neuroendocrine tumors, a standard therapeutic strategy has not been established. Treatment modalities including surgery, chemotherapy, radiotherapy, endoscopic treatment, biological therapy and targeted therapy have been reported [4]. The choice of treatment is then guided by the staging of the tumour.

The combination of radical resection, radical lymph node dissection and chemotherapy produces significantly better results than radiotherapy and chemotherapy alone or in combination, and can prolong survival, improve quality of life and prognosis in patients with limited-stage disease [5-6].

Thanks to improvements in therapeutic digestive endoscopy techniques, endoscopic submucosal dissection can now be offered for esophageal neuroendocrine tumors diagnosed at an early stage [7]. Metastatic forms are treated with palliative chemotherapy or radiotherapy, and symptomatic supportive care [8]

4. Conclusion

Neuroendocrine tumours of the oesophagus are characterized by low incidence, aggressiveness and lack of specific symptoms, leading to delayed diagnosis, which in turn worsens prognosis. The management of these tumors remains a challenge, as there are no official guidelines.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no competing interest.

Statement of ethical approval

Ethical approval was obtained.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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