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Case Report

A Rare Case of a Neuroendocrine Tumor of the Anal Canal: Case Report

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Abstract

We describe a rare case of neuroendocrine tumor of the anal canal with a dismal prognosis, as well as the importance of immunohistochemistry in determining a correct diagnosis and guiding treatment.

Introduction

Squamous cell carcinoma (SCC) and adenocarcinomas are the most common anal cancers, with anal neuroendocrine carcinomas (ANECs) accounting for only 1% of anal malignancies [1].

Small, well-differentiated neuroendocrine tumors are far more prevalent, measuring less than 10 mm in diameter and rarely invading or metastasizing at the time of diagnosis [2]. Ki-67 proliferation index is used

to classify neuroendocrine neoplasms, ranging from G1 (Ki-67 index < 3%), G2 (3-20%), to G3 (> 20%). G3 tumors are further split into two types: well-differentiated NET G3 and non-epithelial carcinomas (NECs) [3].

ANECs can grow in tandem with squamous cell carcinoma [4] or adenocarcinoma [5], although the related pathophysiology is unknown. However, they are not derived from well-differentiated neuroendocrine tumors [6].

Case Description

Anal hemorrhage, small size stool with purulent discharge, and severe proctalgia were described by an 80-year-old woman with a history of high blood pressure under therapy. In the last three months she had no abdominal pain, no bowel habit changes, no fever but a weight loss. A hard anal mass (Fig. 1A) were detected in the rectal exam and the pathological evaluation revealed neoplastic cells that were immunoreactive for cytokeratin (CK35) (Fig. 1B), chromogranin A (CgA) (Fig. 1C) and synaptophysin (SNP) that were compatible with poorly differentiated small cell ANEC with a Ki-67 proliferation index of 90%.

The rest of the assessment revealed a local extension with hepatic and bone metastasis. Following an interdisciplinary consultation meeting, the treatment decision was palliative chemotherapy based on 5 Fluorouracil (5FU) (600 to 1000mg/m^2) from Day 1 to Day 5 and cisplatin (80 to 100mg/m^2) on day 1. Unfortunately she died 3 months later from septic complication.

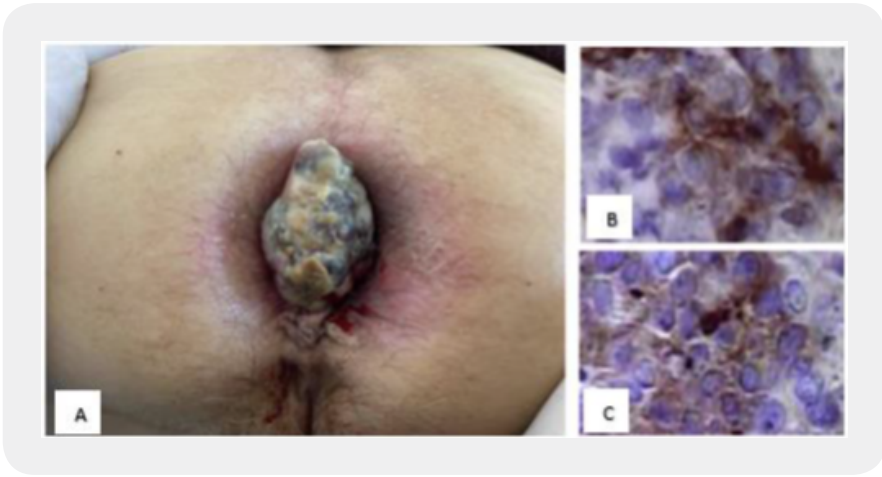

Figure 1: *Tumoral process evolving up to 9cm from the anal margin, not stenosing (A). Immunohistochemical staining was compatible with high grade NET by showing positive neoplastic cells for CK35 (B) and CgA (C).*

Discussion

In this study, a patient with a very proliferative ANEC is described. The presence of an anal tumor is uncommon and predicts a poor prognosis [7,8].

There are several types of NET, rare and aggressive, that differ in multiple ways. The clinical appearance of NET is similar to that of colorectal adenocarcinomas. At the time of diagnosis, however, a more advanced tumor stage can be seen.

Carcinoid (diarrhea and rash) and metabolic problems are rare signs of paraneoplastic illness [9]. Anal adenocarcinoma and anal squamous skin cancer are linked to high-risk papillomavirus [10], and HPV subtypes 16 and 18 have previously been found in ANEC [5,10]. NSE, CD56, CgA, and synaptophysin are particular markers that can be used to determine neuroendocrine differentiation, with the latter two being recommended due to their relative sensitivity and specificity [11].

Surgical excision provides the best chance of cure for people with NET, an immunohistochemical investigation is also important to guide treatment. It is true that complete remission of poorly differentiated NECs is uncommon. The median survival time for metastatic NECs from diverse locations is 7.6 months for lung NEC, 7.5 months for gastrointestinal NEC, and 2.5 months for NEC of unknown source [12].

Because adjuvant therapies such as radiotherapy and chemotherapy do not increase survival in these patients, only early identification of the disease can have some impact on its progression. However, novel chemotherapeutic regimens combining streptozotocin and 5-fluorouracil or doxorubicin and 5-fluorouracil are being developed. Palliative therapies such as surgery, colostomy manufacturing, and radiotherapy should also be considered to improve the quality of life of individuals with advanced disease.

Conclusion

Anus neuroendocrine carcinoma is quite uncommon. To make a diagnosis, immunocytochemistry is required. Chemotherapy is the mainstay of treatment, despite the fact that the response is only temporary. Radiotherapy is mostly used for local control and symptom alleviation. Despite local control, the prognosis is dismal, with common early distant metastases.

Conflict of Interest

The authors have no conflicts of interest to declare.

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