Neuroendocrine Tumor of Esophagus

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Introduction

Esophageal carcinoids remain one of the rarest of all carcinoid tumors, with only 32 cases reported in literature. This kind of tumor exhibits a marked male predominance, with a male-to-female ratio of 6:1.1 The age at diagnosis is widely variable, ranging from 30 to 82 years, and patients present with multiple symptoms, most commonly dysphagia, weight loss, pain and reflux esophagitis.1,2 In only one case the presence of carcinoid syndrome was reported. Interestingly, most esophageal carcinoids are localized in the lower third of the esophagus or at the gastroesophageal junction, paralleling the increasingly distal distribution of endocrine cells.3

Case Report

Identification
Male,
64 years old

Past medical history
Chronic gastritis

History of present illness
Patient with burn symptoms of epigastric region was submitted to gastric endoscopy with biopsy that showed epidermoid carcinoma of the esophagus.

Complementary examination
Endoscopy: esophageal lesion in lower third
TAC: dense gastroesophageal junction. Adenopathy in celiac trunk with 2,3cm (Fig.1a, 1b).
Octreoscan: Normal (Fig. 2a, 2b)

Surgery treatment
The patient was submitted to laparotomy and left cervicotomy to perform esophagectomy with transhiatal esophagogastrotomy.

Histology:
Small cell neuroendocrine carcinoma of the esophagus (Fig. 3a - low amplification 40x) with angioinvasion and metastasis in regional lymph nodes. Subepithelial infiltration by cells with dark nuclei of round on oval shape and very scanty cytoplasm growing in a predominantly diffuse fashion (3b – high amplification 100x). Tumor cells are positive for synaptophysin (Fig. 4) and MNF116 (Fig.5).

Evolution
The patient was discharged 12 days after surgery. A few weeks later he complained of dysphagia and the introduction of endoluminal prosthesis became necessary. Patient died sixth month after surgery.

Conclusion

As evidenced by the idiosyncratic features of carcinoid lesions, increased awareness of such tumors will facilitate preoperative diagnosis and enable the institution of appropriate therapy. As a consequence of misdiagnosis inadequate treatment would occur. The diminution of the likelihood of inadvertently neglecting these often benign, indolent neoplasms that are well known to metastasize if unaddressed, would represent an important advance. Awareness of the possibility that such lesions may exist in unusual sites is an important issue in assuring that definitive therapy is instituted and prognosis is maximally amplified.

References