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Case Report

Bilateral Invasive Orbital Metastases from a Poorly Differentiated Large Cell Neuroendocrine Carcinoma of the Esophagus

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Abstract

We report a case of a 62-year-old man presenting with a right eye tumor and retinal detachment. A magnetic resonance imaging (MRI) of the skull showed hyperintense bilateral orbital masses exhibiting an intraocular component and an extraconal component in the right eye measuring 21.8 x 23.2 mm. Immunohistochemistry studies and histopathology results of our patient following right orbital exenteration confirmed a large cell neuroendocrine carcinoma. A positron emission tomography-computed tomography (PET/CT) scan was performed in an attempt to identify the primary origin detecting a metabolically active tumor located in the distal esophagus and metastases to the bone, liver, pancreas, left kidney, mesentery, abdominal, mediastinal and left axillary lymph nodes, choroid of the left eye and lumbar soft tissues. An upper endoscopy revealed an esophageal infiltrating mass. Immunohistochemical staining confirmed a poorly differentiated, large cell neuroendocrine carcinoma.

Keywords: Large cell neuroendocrine carcinoma, Esophagus, Orbital, Metastasis

Introduction

The prevalence of orbital metastases has increased in the last years and is estimated to range from 2 to 4.7% [1]. Any cancer that can spread through the hematogenous route can metastasize to the orbit and ocular adnexa. The primary cancers that most commonly lead to intraocular metastases include breast cancer, lung cancer, prostate cancer, melanoma and carcinoid tumors [1]. Metastasis from carcinoid tumors tend to become apparent late in the course of malignancy usually from a gastrointestinal site [1-3].

Neuroendocrine tumors (NETs) represent an unusual and complex disease spectrum, which appear to have increased in overall incidence over the past 30 years. The greatest incidence of NETs includes gastrointestinal tract and respiratory system neoplasms. In the gastrointestinal tract, most NETs occur in the small bowel (41.8%), rectum (27.4%), and stomach (8.7%) [4].The presence of NETs in the esophagus is rare but not exotic. Most are poorly differentiated neuroendocrine carcinomas [5-8].

Herein we present a particularly interesting case, to the best of our knowledge, the first case report of orbital metastasis (OM) as the inaugural manifestation of a neuroendocrine carcinoma of the esophagus with widespread metastases.

Summary of Clinical and Pathological Findings

A 62-year-old man, who consulted for a 6-month history of bilateral progressive decrease in vision and acute orbital pain diagnosed as bilateral glaucoma. No other remarkable symptoms were noted. His past history included smoking for 30 years. Ocular examination revealed a right retinal mass with retinal detachment. No mass was evidenced in the contralateral eye. A magnetic resonance imaging (MRI) of the skull showed bilateral hyperintense orbital masses, with an intraocular component (choroid) and extraconal component in the right eye measuring 21.8 x 23.2 mm (Figures 1A and 1B). An optical coherence tomography (OCT) revealed a superior and inferior temporal peripapillary dome shaped lesion, with hypoechoic areas suggestive of necrosis, scleral rupture and orbital infiltration. A 19.2 x 20.3 mm choroidal mass was evidenced in the left eye. The initial clinical impression was bilateral choroidal melanoma with invasive orbital involvement on the right side. Patient underwent right orbital exenteration due to infection of the soft tissues.

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