Image Findings of a Rare Case of Neuroendocrine Tumor Metastatic to Orbital Extraocular Muscle in Gallium-68 DOTANOC Positron Emission Tomography/Computed Tomography and Therapy with Lutetium-177 DOTATATE

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Abstract

Metastatic tumor is one of several etiologies of space-occupying masses in the orbit that accounts for 1-13% of all orbital masses. In the adult patient population, breast cancer is the most common tumor to metastasize to the orbit, followed by metastasis from the lung, prostate, and gastrointestinal tract. Carcinoid tumors are rare neuroendocrine neoplasms derived from enterochromaffin cells, which are found primarily in the gastrointestinal tract and bronchial tree. Liver metastases are the classic presentation of distant disease. Although rare, metastatic carcinoid to the extraocular muscles (EOMs) has been relatively well described in both retrospective case reports and clinical series in the ophthalmology literature, but not in nuclear medicine. Positron emission tomography/computed tomography (PET/CT) using Ga-68-labeled somatostatin-analogues have shown superiority over other modalities for imaging of Neuroendocrine tumor We describe a case of bilateral EOM metastasis from carcinoid lung in Ga-68 DOTANOC PET/CT and treatment with Lu -177 DOTATATE.

Key words: *Extraocular muscle metastasis,Ga-68 DOTANOC, Lu- 177 DOTATATE, neuroendocrine tumor, positron emission tomography/computed tomography*

Introduction

Carcinoid tumors are low grade neoplasms usually arising from neuroendocrine cells of the bronchial and gastrointestinal tracts.[1,2] Although carcinoid tumors metastasize in 50-75% of patients with the most common sites being lymph nodes, liver, and bones, metastases to the extraocular muscles (EOMs) have only rarely been reported.[3] Diplopia and proptosis are the most common presenting symptoms and signs of orbital metastasis. Gallium-68 labeled somatostatin analogues are short peptide analogues of somatostatin which are linked to the positron-emitter Ga-68 by a bifunctional chelate, namely 1,4,7,10-tetraazacyclododecane-1,4,7,10-tetraacetic acid (DOTA) (Ga-68 DOTANOC). We describe rare case of primary lung carcinoid with widespread nodal metastasis and bilateral EOM metastasis seen in Ga-68 DOTANOC positron emission tomography/computed tomography (PET/CT) and treated with lutetium-177 (Lu-177) DOTATATE.

Case Report

A 45-year-old female patient presented with weight loss, mild proptosis, and decreased vision in both the eyes. She was evaluated to have multiple lymphnodes and biopsy from mediastinal lymphnode showed metastatic neuroendocrine tumour. She was referred for whole body Ga-68 DOTANOC PET/CT for detection of primary site. Whole body PET/CT showed an intense uptake in the left lung nodule, mediastinal, and retroperitoneal lymph nodes and also uptake in the EOM lesions (arrows) [Figure 1]. A diagnosis of primary carcinoid lung with nodal and extrocular muscle metastasis was made and she was treated with Lu-177 DOTATATE (7.4GBq). Whole body Lu-177 images [Figure 2] showed intense uptake in lymphnodes and bilateral EOM metastasis (arrows). Symptomatically, she had improvement with weight gain and normal vision in both eyes on clinical examination.

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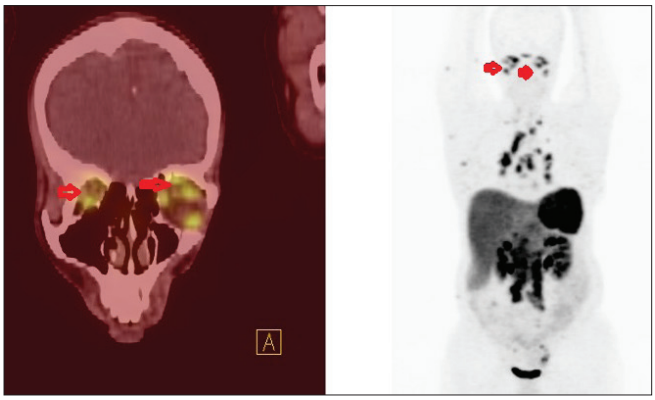

Figure 1: Whole body gallium-68 DOTANOC-PET/CT coronal fused and maximum intensity projection image showing intense uptake in the bilateral extraocular soft tissue muscle metastasis (arrows). Also, multiple mediastinal and retroperitoneal lymphnodes noted

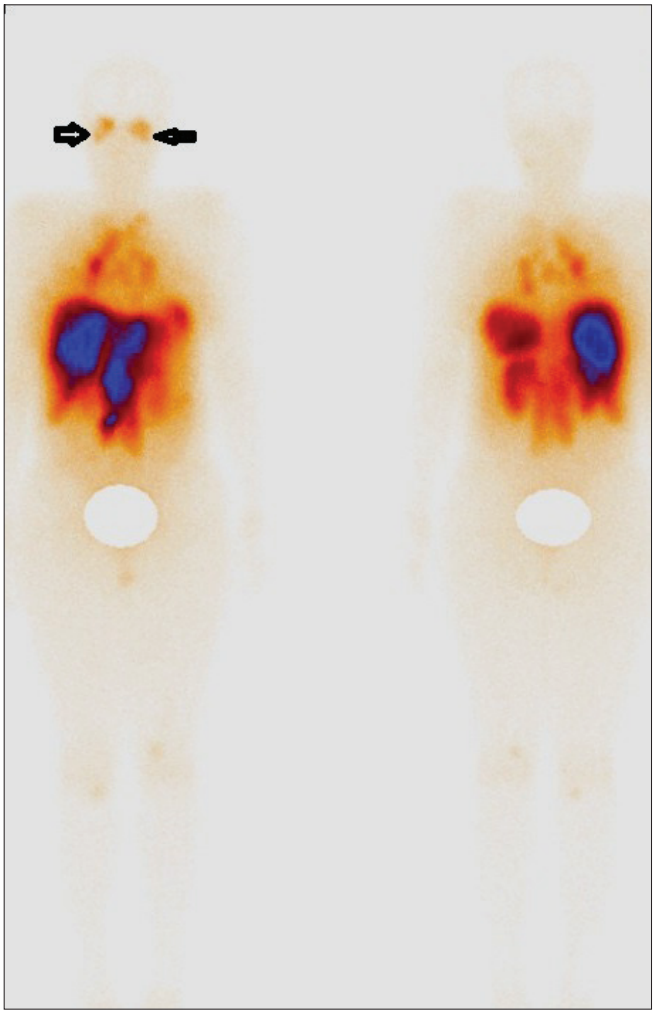

Figure 2: Whole body Lu-177 DOTATATE images showed intense uptake in lymphnodes and EOM metastasis

Discussion

Neuroendocrine tumors are slow growing, low grade neoplasms that usually develop in the gastrointestinal tract and bronchus but can rarely originate in the ovary, thymus, parotid gland, breast, and testis.[1,2] Although they are thought to be of low malignant potential, they can metastasize to various sites such as liver, lymph nodes, and bone. Neuroendocrine tumor metastasis to the liver can lead to carcinoid syndrome due to release of bioactive amines from the tumor. Neuroendocrine tumor metastasis to the eye and orbit is rare.[3] Goldberg *et al*.[4] reported 11 cases of carcinoid tumor metastasis to the orbit out of 245 orbital lesions and discussed the various presenting symptoms and signs and the pathological findings of the lesions. Diplopia and proptosis are the most common presenting symptoms and signs of orbital metastasis. Regarding bilateral and multifocal disease, a study by Mehta *et al*.[5] of 13 patients with orbital carcinoid metastases showed that only 2 of 13 patients had bilateral lesions and no quadrant was favored. Our patient had proptosis, decreased vision, and bilateral involvement. Metastatic orbital tumors are usually isointense to EOMs and hypointense to orbital fat on T1 images and mildly hyperintense to EOM and fat on T2 images with enhancement following contrast administration in magnetic resonance imaging.[6]

Metastatic neuroendocrine orbital tumors have been reported to have increased iodine-131 metaiodobenzylguanidine (MIBG) uptake attributed to the elevated serotonin levels in these tumors.[7] Isidori *et al*.[8] have evaluated the use of I-131 MIBG and indium octreotide scanning in 40 cases of neuroendocrine tumors including six patients (15%) with ocular metastasis (five choroidal lesions and one orbital lesion). None of their patients showed positive uptake with the I-131 MIBG, but four of the six showed positive uptake with Indium scintigraphy. We used recent modality for NET, Ga 68 DOTANOC PET/CT and still now no case is reported with EOM metastasis from NET. Treatment options for orbital neuroendocrine tumor metastasis include excision, orbital exenteration, radiotherapy, hormonal therapy, and chemotherapy.

A review by Makis *et al*.[9] of 251 neuroendocrine tumors treated with Lu-177 DOTATATE or I-131 MIBG since 2003 revealed four patients with orbital metastases (1.6%), two treated with Lu-177 DOTATATE, and two with I-131 MIBG. Of these four patients, one patient was symptomatic with diplopia and eye pain, and two patients had physical signs of orbital involvement. The symptomatic orbital metastasis improved with Lu-177 DOTATATE therapy, and proptosis improved with I-131 MIBG therapy in one of two patients. We treated our patient with Lu-177 DOTATATE and she had improvement in vision. Given that patients with carcinoid tumors may have prolonged survival despite dissemination, maintaining quality of life by providing early diagnosis and effective treatment to preserve vision and comfort is a fundamental issue. Our case describes the findings of Ga 68 DOTANOC PET CT and treatment with Lu 177 DOTATATE in metastatic carcinoid to orbit.

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Conflicts of interest

There are no conflicts of interest.

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