

Sinonasal Malignant Melanoma with Diffuse Carcinomatosis: Initial PET/CT Staging and Follow-up CT/MR Imaging

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Mucosal malignant melanoma of the head and neck is a rare but aggressive type of melanoma with poor clinical prognosis and outcome. Its nonspecific symptomatology frequently leads to its delayed diagnosis until advanced stages of the disease. The author presents a case of primary sinonasal amelanotic melanoma with preoperative F-18 FDG PET/CT showing diffuse bone metastasis only two months after the onset of epistaxis and nasal congestion, and subsequent postsurgical CT/MR showing diffuse soft tissue tumor dissemination and malignant pleural effusion.

Introduction

Sinonasal melanoma is an uncommon malignancy difficult to diagnose due to its hidden locations of the head and neck and due to its deceptive symptoms. Contrary to its cutaneous counterpart, sinonasal melanoma has a high propensity for metastasis and a poor survival rate despite aggressive therapy. To illustrate the unpredictable behavior of such neoplasm, the author presents a case of advanced disease at initial diagnosis with accelerated hepatic, pleural, peritoneal and retroperitoneal dissemination.

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Abbreviations: CT, computed tomography; MR, magnetic resonance; PET, positron emission tomography; SMM, sinonasal malignant melanoma

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Case Report

A 56-year-old woman came to our institution with a 2-month history of nasal congestion and epistaxis. Otorhinologic consultation found a left nasal mass, the biopsy of which yielded histologic evidence of primary sinonasal malignant melanoma (SMM). Head and neck MR examination showed a hypervascular lesion of the left nasal cavity extending to the anterior left ethmoid air cells and frontal sinuses. There was no orbital or intracranial tumor involvement. A preoperative F-18 FDG PET/CT staging study confirmed the hypermetabolic left SMM (Figure 1). It also showed diffuse foci of abnormal radiotracer uptake in the axial and appendicular skeleton compatible with diffuse osseous metastasis (Figure 2). Shortly after the PET/CT imaging, the patient underwent tumor resection with left lateral rhinotomy and maxillectomy. The surgical specimen was consistent with a left amelanotic SMM with extensive ulceration and no host response. The tumor showed about 20 mitotic figures per mm² with strong positivity for HMB 45 and Melan-A. There was lymphovascular and perineural tumor involvement.

Sinonasal Malignant Melanoma with Diffuse Carcinomatosis: Initial PET/CT Staging and Follow-up CT/MR Imaging

Spine MR examination, performed three weeks after PET/CT, showed diffuse bone marrow signal intensity changes from osseous tumor infiltration and detected new prevertebral adenopathy not seen on initial PET/CT (Figure 3). Ensuing iliac crest biopsy showed near total effacement of normal marrow, which was replaced by tumor cells identical to the ones from the sinonasal mass. CT, following the MR examination, showed diffuse tumor dissemination to lungs, liver, mesentery, peritoneum and retroperitoneum, and bilateral malignant pleural effusion (figure 4). These features of advanced carcinomatosis were not conspicuous on the preoperative PET or co-registered multi-detector CT staging examination. The patient developed progressive clinical deterioration with hypoxia, anasarca and pancytopenia from diffuse bone marrow tumor infiltration. She passed away less than two months after SMM surgery.

Discussion

SMM is an uncommon tumor accounting for 0.3-2% of all malignant melanomas, 4% of head and neck melanomas and 4% of all sinonasal neoplasms [1]. At least one

third of SMM are amelanotic [2, 3]. SMM has equal gender distribution targeting a patient population of 6 decades of life [1, 4]. It arises predominantly from nasal cavity or nasal cavity and sinuses [5]. The nonspecific symptoms, which include epistaxis, nasal congestion and obstruction, nasal discharge and pain, may prolong the duration between SMM onset and its definitive diagnosis. Diplopia, proptosis and neurological symptomatology are usually findings of advanced stages of the disease with orbital and intracranial invasion. SMM has a high propensity to disseminate due to the rich lymphatic and vascular network of the sinonasal tract. The metastatic status is the most important factor for prognosis and outcome of the disease [1]. Additional prognostic factors include patient's age, mitotic characteristics of the melanoma, tumor thickness, involvement of the nasopharynx, nasal obstruction, vascular invasion and tumor recurrence [1, 6, 7]. SMM is a very aggressive malignancy with 5-year and 10-year survival rates ranging from 10 to 47% and 20 to 24.3% respectively [6]. CT and MR are the main imaging modalities for detailed anatomic evaluation of local tumor involvement [2]. MR provides different patterns of tumor intensity changes on T1 and T2-weighted sequences depending on

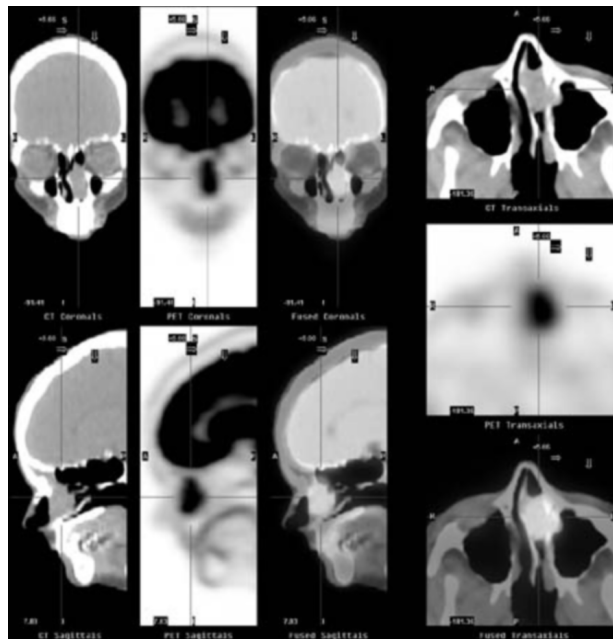


Figure 1. 56-year-old woman with sinonasal amelanotic melanoma. Composite PET/CT image (sequence: CT, PET, Fused PET/CT in the coronal, sagittal and axial projections) shows a high metabolic lesion within the left nasal cavity extending to the left ethmoid sinuses (crosshair).

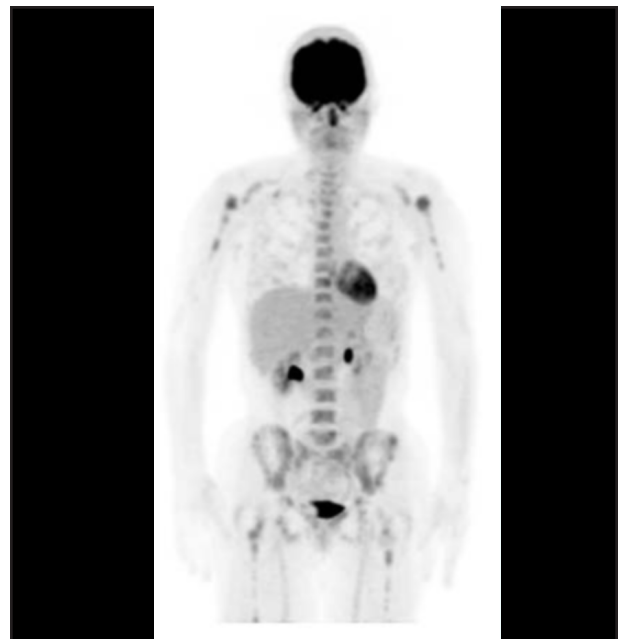


Figure 2. Anterior MIP PET image shows, in addition to the left sinonasal melanoma, diffuse foci of abnormal F-18 FDG uptake at the entire spine, shoulder and pelvic girdles, and long bones of extremities related to diffuse osseous metastases.

Sinonasal Malignant Melanoma with Diffuse Carcinomatosis: Initial PET/CT Staging and Follow-up CT/MR Imaging



Figure 3. Sagittal T1-weighted MR image of the thoracic spine, obtained 3 weeks after sinonasal surgery, shows diffuse abnormally decreased signal intensity of the bone marrow comparing to the signal intensity of adjacent vertebral disks. There is prevertebral and retrocrustral adenopathy (arrows), which was not present on initial PET/CT examination.



Figure 4. Axial CT image of the abdomen, obtained 3 weeks after sinonasal surgery, shows diffuse metastases involving the liver, peritoneum and retroperitoneum (arrows).

the melanotic or amelanotic constitution of SMM [8]. Comparing to conventional cross-sectional imaging, F-18 FDG PET/CT provides a more comprehensive whole-body assessment thus is more able to detect regional nodal and distant metastasis, which could be present at the initial presentation of the disease [9-12]. PET is based on high glucose metabolism of melanoma and may not distinguish a melanotic variant from an amelanotic one. Treatment consists of aggressive surgery for local control of SMM. Complete resection of the malignancy may difficult due to involvement of adjacent vital structures of the head and neck. Post-operative radiation therapy may have limited positive impact on the survival rate [13, 14]. Chemo-immunotherapy still needs clinical validation [6].

In this patient, the accelerated tumor dissemination in the immediate postoperative course may be related to combining factors of aggressive sinonasal melanoma behavior, markedly depressed immune system of the patient and traumatic maxillofacial surgery releasing malignant cells into the systemic circulation. Alternatively, the diffuse soft tissue metastasis may exist at microscopic level by

the time of the PET/CT staging beyond the resolution of this modality. PET has known limitation for evaluating small tumors and for assessing subtle liver lesions due to physiologic background tracer uptake of this organ [15, 16]. The initial PET staging with its co-registered multi-detector CT did not show any obvious liver metastasis or peritoneal/mesenteric carcinomatosis. These lesions may rapidly progress in this immunodepressed patient after major head and neck surgery [17].

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**Sinonasal Malignant Melanoma with Diffuse Carcinomatosis:
Initial PET/CT Staging and Follow-up CT/MR Imaging**

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