Abstract

Objectives: Esthesioneuroblastoma (ENB) is a rare neoplasm arising from the olfactory epithelium of the upper nasal cavity. It exhibits various clinical behavior with the potential for locoregional recurrence and metastasis. The utility of PET/CT in the evaluation, staging, and follow-up of patients with ENB has not been well characterized, nor has the relationship between PET/CT findings and histologic grade. This study aims to evaluate the potential clinical utility of PET/CT in predicting the tumor’s histologic grade.

Study design: Retrospective chart review.

Methods: Seven patients treated for ENB who also had a PET/CT scan were included in the study. All patients had PET/CT scans 1-6 weeks before surgery, and lesions were assigned a peak SUV (standardized uptake value) that was standardized to the liver uptake. Pathology specimens were categorized into grades I-IV according to Hyams histologic grading criteria. The ratio of the lesion to liver uptake was then plotted against the pathological grade of the surgical specimen.

Results: Six out of seven patients had direct correlation between the degree of lesion uptake on PET/CT and Hyams histologic grade. Recent data in the literature notes that the Hyams staging system may be an important factor in determining prognosis for tumor recurrence and survival. Although a small series, our study suggests that PET/CT may be utilized to predict the tumor’s histologic grade. Lesion uptake on PET/CT may be utilized as a predictor of outcome and the need for post-surgical adjunctive treatment.

Introduction

Esthesioneuroblastoma (ENB) or olfactory neuroblastoma is an uncommon malignancy of the nasal cavity derived from the olfactory epithelium. It comprises 3-5% of nasal and paranasal sinus neoplasms. Due to its high origin in the nasal vault from the superior one third of the nasal septum, cribriform plate, and superior turbinates, ENB can present with locally advanced disease involving anterior cranial fossa and paranasal sinuses. Due to the rarity of this neoplasm, data on clinical behavior, prognostic factors, optimal treatment modality, and post-treatment surveillance for recurrence is limited. Hyams’ histologic grading system, which is most commonly used, is based on a statistical significant prognostic factor for many recent studies, correlating with disease survival and recurrence using PET. Hyams histologic grading system was developed in 1988, and classifies ENB into 4 grades based on native architecture preservation, mitotic index, nuclear polymorphism, matrix characteristics, the presence of rosettes, and tumor necrosis. Our study aims to evaluate the potential clinical utility of PET/CT in predicting the tumor’s histologic grade, as FDG uptake was found to correlate directly with degree of histologic grade by Hyams criteria in nearly all of our patients. The utility of PET/CT in pre and post-treatment surveillance for ENB is not well defined, and this study highlights its potential role.

Methods

Seven patients treated for ENB who also had a PET/CT scan were included in the study. All patients had PET/CT scans 1-6 weeks before surgery, and lesions were assigned a peak SUV (standardized uptake value) that was standardized to the liver uptake. Pathology specimens were categorized into grades I-IV according to Hyams histologic grading criteria. The ratio of the lesion to liver uptake was then plotted against the pathological grade of the surgical specimen.

Results

Six out of seven patients had direct correlation between the degree of lesion uptake on PET/CT and Hyams histologic grade. Higher SUVs corresponded with higher grade Hyams histology. The one patient with no correlation had a moderate SUV and grade 1 ENB on nasal biopsy, but Hyams grading of surgical resection specimen two months later was a grade 2/3. This latter pair would have correlated the degree of uptake with histologic grade.

Discussion

Esthesioneuroblastoma is a rare malignant tumor of olfactory epithelial origin, arising from the superior aspect of the nasal vault. It is a challenging disease due to its anatomic locations, rates of recurrence, regional and distant metastases despite aggressive multimodality treatment. No universal staging system exists, but however the Kadish system is the most commonly used. Kadish divides tumors into 3 groups: group A lesions limited to nasal cavity, group B involve the nasal cavity and paranasal sinuses, and group C extend beyond the nasal cavity and paranasal sinuses. Although most frequently used, some critique this staging system as nonspecific and all inclusive, not accounting for exact locations of tumor involvement. Morita et al expanded this latter staging system, adding a group D for regional or distant metastasis, although this is less frequently utilized. Age distribution is typically unimodal, with mean age in the 50s and the majority of patients between 40 and 70 years of age, although a bi-modal distribution has also been described.

Many patients with advanced stage disease due to the insidious and nonspecific nature of presenting symptoms: 66% with nasal obstruction, 60% with recurrent epistaxis, 12% headaches, 12% rhinorrhea, 8% visual disturbances, 6% anosmia, 6% proptosis and 3% swelling in cheeks. A meta-analysis of 26 studies between 1990 and 2000 revealed a 45% 5-year overall survival compared with 62% based on SEER data. Kadish staging criteria has been proposed as a major prognostic factor, with 89% and 83% respective 3-year survival in patients with stages A & B, and 53% with stage C. Hyams histologic grading has great prognostic value, with advanced Hyams grade tumors having a significantly worse prognosis despite aggressive multimodality treatment. Presence of cervical lymphadenopathy has also been posed as a major prognostic factor, raising the question of whether N0 necks should be electively treated. Treatment options have included extracranial surgical resection, craniospinal radiation, chemotherapy or a combination, although the exact indications for adjunctive treatment have not been well elucidated.

This study correlates histologic grade of ENB with FDG avidity on PET/CT. Although a small series, our study suggests that PET/CT may be utilized to predict the tumor’s histologic grade. Lesion uptake on PET/CT may be utilized as a predictor of outcome and the need for post-surgical adjunctive treatment. It also highlights the potential role of PET/CT in post-treatment surveillance for recurrence and regional and distant metastases.

Case series

Case 1: 44-year-old male with newly diagnosed ENB underwent staging PET/CT with which demonstrated moderate increased FDG uptake in the left nasal cavity mass (red solid arrow), the uptake extended into the left ethmoid sinus. Patient subsequently underwent anterior craniofacial resection and medial maxillectomy. Surgical pathology demonstrated Grade I ENB.

Case 2: 46-year-old male who presented with newly diagnosed ENB in the right orbit underwent staging PET CT scan which demonstrated a focus of intense FDG uptake (solid red arrow) at the inner canthus of right eye. Patient subsequently underwent right lateral rhinotomy and orbitotomy with removal of nasal/medial maxilla and orbital tumor.

Transaxial, coronal and sagittal CT PET fused images demonstrate highly FDG avid lesion at the inner canthus of the right eye.

References