Case Report

Role of F18 fluorodeoxyglucose positron-emission tomography/computed tomography in the management of Askin's tumor

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ABSTRACT

A primitive neuroectodermal tumor (PNET) of the thoraco-abdominal region is one of a group of small round cell tumors usually found in children and young adults, originally described by Askin *et al*. Most cases arise in the soft-tissues of the thorax, but may rarely occur within the lung with the symptoms of chest wall pain, pleural effusion and dyspnea. The authors present two cases demonstrating the utility of F18 fluorodeoxyglucose positron-emission tomography/computed tomography in the staging and prognosis of PNET of the chest wall.

Keywords: Chest wall, F18 fluorodeoxyglucose, positron-emission tomography/computed tomography, primitive neuroectodermal tumor

INTRODUCTION

A primitive neuroectodermal tumor (PNET) of the thoraco-abdominal region is one of a group of small round cell tumors usually found in children and young adults. It was originally described by Askin *et al*.,[1] and is associated with a chromosome 22 translocation. While most cases arise in the soft-tissues of the thorax, they may rarely occur within the lung.[2] The most common presenting symptom is chest wall pain, which can be accompanied by dyspnea and pleural effusion. The following cases demonstrate the utility of F18 fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) in the staging and prognosis of PNET of the chest wall.

CASE REPORTS

Case 1

A 35-year-old male patient was diagnosed with a PNET of the left chest wall for which he was treated with 15 cycles of chemotherapy. As there was progressive swelling in the left chest wall over 5 months, he was referred for restaging with FDG PET/CT. Increased FDG uptake (maximum standardized uptake value [SUV_{max} 6.7]) was noted in multiple soft-tissue masses arising from the left chest wall [Figure 1a-c]. The largest (measuring 17.4 cm × 9.9 cm) in the lower half of the left hemithorax showed photopenic regions suggestive of necrosis. Erosion of the left 10th and 11th ribs was also noted, along with satellite mass lesions. Since, the scan was suggestive of residual disease, the patient was further treated with five more cycles of chemotherapy in view of the residual disease. Two months after the last cycle of chemotherapy, a follow-up PET/CT showed increase in size of the lesions with extrathoracic and intra-abdominal extensions [Figure 1d-f]. However, there was no change in the FDG avidity (SUV_{max} 6.6) indicating metabolically stable disease. The patient expired 3 months later.

Case 2

A 14-year-old boy presented with a history of pain in the right chest for 2 months. Fine-needle aspiration from the lesion was consistent with PNET and he was referred for F-18 FDG PET/CT for initial staging of the disease. Intense FDG uptake (SUV_{max} 12.6) was seen in a circumscribed heterogeneously enhancing soft-tissue mass (measuring 13.0 cm × 11.7 cm × 8.0 cm) in the right hemithorax, arising from the chest wall [Figure 2a-d]. Areas of photopenia were seen within the mass, suggestive of necrosis. The adjacent posterior part of the 6th rib was directly infiltrated by the lesion.

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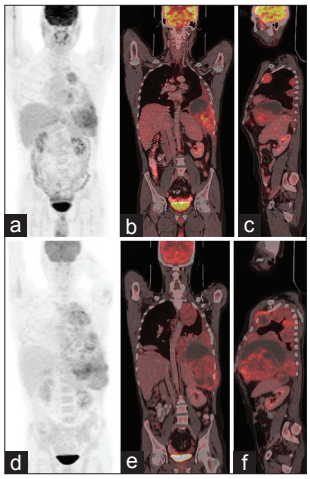

Figure 1: (a) Maximum intensity projection image of initial positron-emission tomography/computed tomography showing abnormal fluorodeoxyglucose uptake in the left chest (b) coronal PET/CT showing FDG avid mass in the left hemithorax with photopenic regions corresponding to hypodensity suggestive of necrosis (c) sagittal PET/CT showing satellite lesions along the left 2nd rib anteriorly and at the costo-vertebral junctions of the left second and 5th ribs (d) MIP image (follow-up PET/CT) showing FDG uptake in the left chest and hypochondriac regions (e) coronal fused PET/CT showing an increase in size of the primary soft-tissue mass (f) sagittal fused PET/CT showing an increase in size of the satellite lesions

Tracer uptake was also seen in the metaphyseal region of the right humerus and left 2nd rib with mild sclerotic changes (not shown), suggestive of metastases. The patient did not respond to chemotherapy and expired after 9 months.

DISCUSSION

PNET of the chest wall or Askin's tumor is a malignant tumor of the Ewing family of tumors comprising small, undifferentiated neuroectodermal cells. The Ewing family of tumors also includes Ewing's sarcoma of the bone, extraosseous Ewing's sarcoma and peripheral neuroepithelioma. These rare tumors are considered to arise from a common origin, the neuroectoderm, in which malignant cells are found in the bone and soft-tissues. They occur most frequently in teenagers. The imaging evaluation of a PNET of the chest wall requires a multimodality approach involving chest radiography, CT and magnetic resonance imaging.[1] FDG PET/CT has been suggested as a complement to CT in the evaluation of sarcomas.[2,3] While this technique can discriminate between low and high grade sarcomas with accuracy of 94% during initial staging, the sensitivity for Ewing's sarcoma has been found to be 100%.[4] However, the role of FDG PET/CT in the management of PNET of the chest wall has not been clearly established. There are several reports describing FDG uptake in PNET,[5-7] but few had PNET of the chest wall.[8-11]

Biologic heterogeneity (including proliferation, necrosis, non-cellular accumulations, differences in blood flow, cellular metabolism, oxygenation, etc.) is a very important feature of malignant tumors. Eary *et al.*,[12] found that SUV_{max} and heterogeneous distribution of FDG can differentiate patients with sarcoma into higher risk and lower risk with different

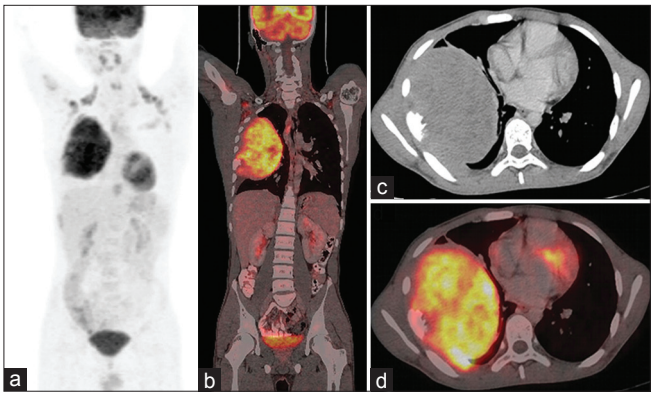

Figure 2: (a) Maximum intensity projection image showing fluorodeoxyglucose uptake in the right chest and brown adipose tissue (arrows) (b) coronal positron-emission tomography/computed tomography showing intense FDG uptake in a circumscribed heterogeneously enhancing soft-tissue mass with photopenic areas suggestive of necrosis, with compression/passive collapse of lung parenchyma. Mild FDG uptake is seen in the metaphyseal region of right humerus (arrow), with mild sclerotic changes (c) axial contrast enhanced computed tomography (d) axial PET/CT showing irregular sclerosis of the cortical margin of the right 6th rib

prognoses. Both patients in our series had heterogeneous FDG distribution with pre-therapy SUV_{max} of 6.7 and 12.6 respectively and this correlates well with the poor prognosis in these patients. In high-grade soft-tissue sarcomas (STS), Evilevitch *et al.*,[13] found that a 60% decrease in tumor FDG uptake following neoadjuvant chemotherapy, can predict histopathological response with 100% sensitivity and 71% specificity. In the first patient, there was hardly any decrease in the FDG uptake following chemotherapy (metabolically stable disease in the second PET/CT). This may explain the poor pathological response observed in this patient, with progression-free survival of 12 weeks from the time of the second PET/CT. Völker *et al.*,[14] evaluated 46 pediatric sarcomas and found that PET was superior in assessing lymph node involvement and bone manifestations. Similarly, FDG PET was able to identify the metastatic lesions in patient 2, which would have otherwise been missed by CT. Although these lesions could have been identified on the bone scan, FDG PET/CT appears useful as a comprehensive modality to evaluate all systems in the staging of a PNET.

In concordance with other studies of STS, our findings in these two patients with PNET indicate that FDG PET/CT appears to be useful in predicting patient prognosis, in assessing response to neoadjuvant chemotherapy and in staging or restaging the disease.

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