

CASE REPORT

Primary Pulmonary Primitive Neuroectodermal Tumour: Potential Role of F18-FDG PET/CT as an Imaging Biomarker

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ABSTRACT

Primitive neuroectodermal tumours (PNETs) occurring directly from the lung parenchyma without involvement of the chest wall or pleura are particularly unusual. We describe a 16-year-old male who presented with severe chest and back pain following a fall during high jump competition. Incidental finding of an opacity at the right lung upper lobe was seen on his chest radiograph. Computed tomography of the thorax demonstrated a lobulated hypodense lesion in the anterior segment of right upper lobe and subsequent biopsy confirmed the diagnosis of Ewing Sarcoma/PNET following histologic and immunohistochemical examination. Whole-body positron emission computed tomography/computed tomography (PET/CT) revealed mild, homogenous F18-fluorodeoxyglucose (FDG) uptake in the lung lesion with no evidence of metastasis. He eventually underwent right thoracotomy and right upper lobectomy. Indeed, F18-FDG PET/CT is a valuable imaging method in demonstrating active focus of glucose metabolism in lung parenchymal lesion, for preoperative evaluation and as baseline scan for patient's monitoring.

Keywords: Primitive neuroectodermal tumour, Pulmonary, Positron emission computed tomography/computed tomography, F18-fluorodeoxyglucose

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INTRODUCTION

Primitive neuroectodermal tumour (PNET) belongs to Ewing sarcoma family and is considered as malignant tumour of undifferentiated and small neuroectodermal cells. Typically, Ewing sarcomas develop in bone structure and soft tissue in adolescents, and predominantly occur in males (1). PNETs occurring directly from the lung parenchyma without involvement of the chest wall or pleura are particularly unusual although aggressive in nature. We describe a 16-year-old male who had an incidental finding of a right lung PNET on chest radiograph after developing severe chest and back pain following a fall during high jump competition.

CASE REPORT

A 16-year-old male presented in March 2016 with severe chest and back pain following a fall during high jump competition. A chest radiograph was performed due to the persistent pain, revealing an incidental finding of a right upper lobe lung opacity (Fig. 1). Thoracolumbar spine radiograph was unremarkable. Computed tomography (CT) of the thorax in early May 2016

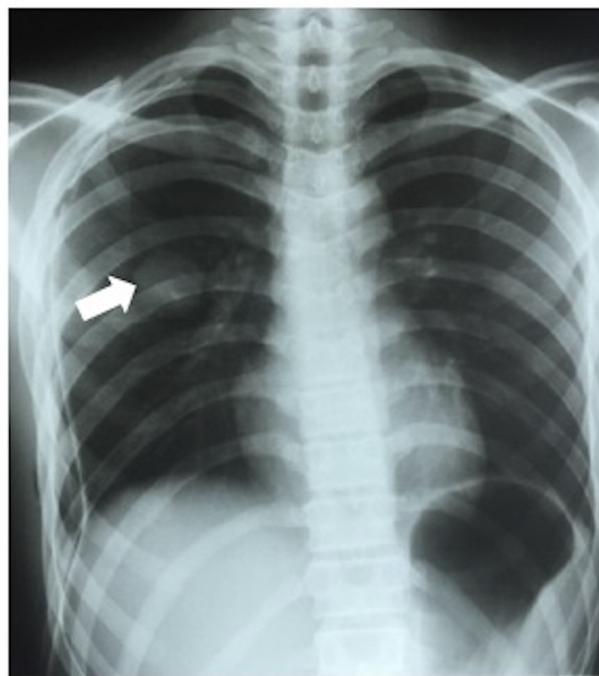


Figure 1: Chest radiograph showing right upper lobe lung opacity (white arrow)

showed evidence of a hypodense lesion with lobulated appearance measuring approximately 2.5 x 2.8 x 2.6 cm in size, in the anterior segment of right upper lobe (Fig. 2). There was no mediastinal lymphadenopathy, pleural effusion or suspicious bony lesion seen. Subsequent

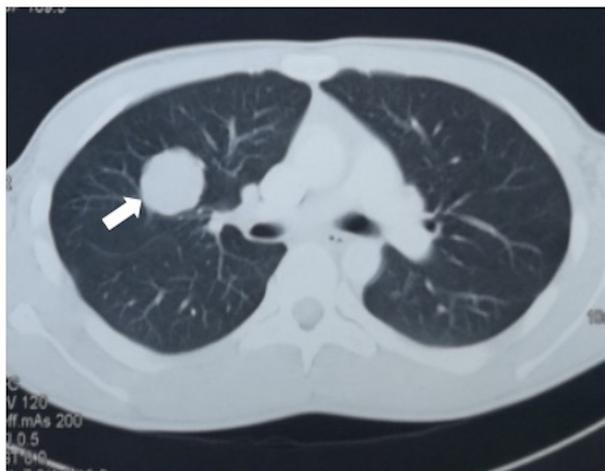


Figure 2: Computed tomography of the thorax revealed a well-defined hypodense lesion in the anterior segment of right upper lobe measuring 2.5 x 2.8 x 2.6 cm (white arrow)

bronchoscopy examination revealed normal findings with no obvious endobronchial mass. Bronchoalveolar lavage for cytology, acid fast bacilli, culture and sensitivity were negative.

Right thoracotomy and open biopsy of the right upper lobe lung lesion was performed few weeks later, confirming the diagnosis of PNET following histologic and immunohistochemical examination (vimentin, EMA, CKAE1/AE3, CD99, NSE and FL11 with scattered CD34 positivity). Whole-body positron emission computed tomography/computed tomography (PET/CT) for tumour staging revealed homogenous F18-fluorodeoxyglucose (FDG) uptake in the lung lesion (standard uptake value, SUV of 2.1) (Fig. 3) with no evidence of FDG avid locoregional, nodal and distant metastasis. Following this, he eventually underwent right thoracotomy and right upper lobectomy procedure in January 2017 and was discharged well. The patient has been regularly followed up until the present at the cardiothoracic and oncology departments as outpatient with no sign of recurrence being observed.

DISCUSSION

PNET belongs to Ewing Sarcomas Family of Tumour, which include Ewing bone tumour (Ewing sarcoma), extraosseous Ewing sarcoma (affecting soft tissues), peripheral neuroepithelioma, Askin tumour (Ewing sarcoma of the chest wall) and atypical Ewing sarcoma. PNETs and Ewing sarcoma appear to be identical not only clinically but also histologically, and commonly emerge during children and teenage years. (1). PNET commonly affects the chest wall or lung in the thoracic region, as a distant metastasis from extrathoracic soft tissue sarcoma. However only few cases of primary pulmonary PNET were being documented (2). In this patient, the tumour was seen at the anterior segment of right upper lobe, confining to the lung with no obvious involvement of the chest wall.

The capability to diagnose PNET has significantly developed with the introduction of an array of immunohistochemical markers, with most tumours to be positively stained on CD99 (84%) and FL11 (90%) (2), as similarly seen in this case whereby the neoplastic cells are positively stained for both CD99 and FL11.

It is indeed essential to exclude the likelihood of lung metastasis from any extrathoracic primary PNET (3). Hence, the imaging evaluation of thoracic region PNET involves a multimodality approach. Plain chest radiography should be the first imaging modality to be performed as it is readily available in most centres. However, CT scan thorax gives better image for the diagnosis of possible involvement of the lungs and ribs, and even metastatic lesions. Pulmonary PNET usually reveals involvement of lung parenchymal only as in this case. Magnetic resonance imaging (MRI) has the capability to provide superior soft tissue contrast and therefore, preferably used in the presence of a large chest wall mass. However, it is quite a challenge to differentiate between malignant and benign soft tissue lesions by relying only on CT scan and MRI examinations (4). The hybrid method, CT in association with PET has

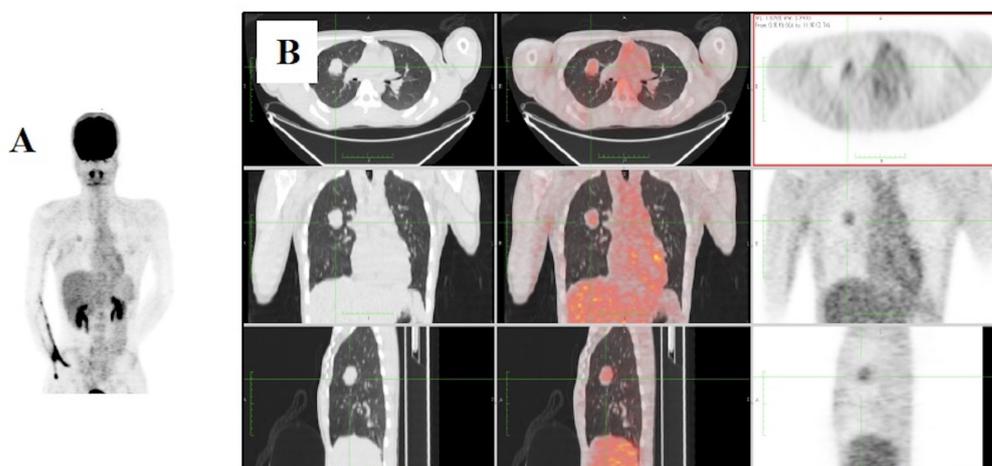


Figure 3A-B: Positron emission computed tomography/computed tomography for tumour staging demonstrated homogenous F18-fluorodeoxyglucose (FDG) uptake in the lung lesion (standard uptake value, SUV of 2.1) with no evidence of FDG avid locoregional, nodal and distant metastasis

been utilised as a powerful tool. It has high sensitivity and specificity as it provides detailed anatomy of lesions and glucose metabolism evaluation. The value of F18-FDG uptake by PNET could be used in staging of the disease, which may be beneficial in predicting the prognosis of the patients and response to the treatment (5). SUV is expressed as the ratio of tissue's activity per milliliter to the activity in the injected dose, whereby it is corrected for decay and patient's body weight, body surface area or lean body mass (4). SUV measurements has the capability to distinguish between benign and malignant lesions as most of the intermediate or high grade soft tissue sarcomas have SUV of more than 2.0, which correlated well with this case. The SUV values of sarcoma has been used to predict the response to neoadjuvant therapy, and being a significant predictor of survival. In addition, PET-CT and other conventional imaging modalities are able to detect almost 100% of primary tumour. However, previous studies have emphasised the advantages of PET-CT as compared to other conventional methods in detecting metastases to bone and lymph nodes (5).

Once the diagnosis is made, multimodality treatment should be started depending on disease status and patient's condition. The management of PNET should be various combination of early surgical interventions, chemotherapy and radiation therapy (1). PNET is an aggressive neoplasm and there have been reports that even with multiple treatments, the two years overall survival is poor with the survival rate of less than 40% for patients with lung metastasis from extrathoracic PNET. However in this case, the patient is currently asymptomatic and well without any chemotherapy and radiotherapy treatment post operation. There was no sign of recurrence during his follow-ups until now.

CONCLUSION

This case emphasises the role of F18-FDG PET/CT as a valuable scanning method to demonstrate active focus of glucose metabolism in lung parenchymal lesion, for preoperative evaluation and as a baseline scan for patient's monitoring.

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