Case Report

Askin's Tumor a Rare Clinical Entity of Thoracopulmonary Region Cresenting as a Case of Paraparesis: Findings on ^{18}F-FDG PET/CT

Abstract

Askin's tumors are rare malignant neoplasms located in the thoracopulmonary region and mainly occur in children and adolescents. In this report, we describe a case of histologically proven Askin's tumor in a 24-year-old male. The patient was admitted with a history of 3-month lower back pain and with a rare presentation of paraparesis.

Keywords: *Askin's tumor, Ewing's sarcoma family of tumors, fluorodeoxyglucose positron emission tomography/computed tomography, fluorodeoxyglucose, paraparesis, peripheral primitive neuroectodermal tumor*

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Introduction

Askin Rosai Tumour/ Askin's Tumour (AT) is a rare primitive neuroectodermal tumor (PNET) developing from the soft tissues of the thoracopulmonary region and primarily occurs in adolescents and young adults aged between 10 and 30 years.[1] It belongs to the Ewing's sarcoma family of tumors sharing the same karyotype abnormality with a translocation involving chromosomes 11 and 22.[2] The most common clinical presentation is a rapidly growing palpable mass overlying the chest wall.[3] Other common clinical presentations are pleural effusion, chest pain, prolonged fever, cough, dyspnea, and hemoptysis. AT is usually localized to the site of origin in the thoracic area at the time of presentation, although the tumor has a tendency to metastasize to regional and distant sites.[3] Common sites of metastases are the lung, bone, bone marrow, and liver. Askin's tumor presenting as compressive myelopathy is extremely rare. Herein, we describe a case of Askin's tumor involving the spine with a rare presentation of paraparesis.

Case Report

A 24-year-old male with no comorbidities presented to the outpatient department with complaints of gradually progressive swelling in the upper back and weakness in the bilateral lower limb for 3 months. At the time of presentation, the patient was paraplegic with 20%–40% sensory loss below D8 vertebral level bilaterally and with urinary and bladder incontinence. Neurological examination revealed a power grading of 5/5 in both upper limbs and 0/5 in bilateral lower limbs. Deep tendon reflex was exaggerated in both lower limbs. The patient was completely bedridden; the bladder was catheterized with Grade 3 bed sores over the buttock region. Physical examination revealed a 4 cm × 6 cm firm lesion in the back region. His magnetic resonance imaging dorsal spine confirmed the presence of a well-defined multilobulated mass lesion epicentered in the right posterior chest wall extending from D5-D10 vertebral level with invasion into the right thoracic cavity causing passive atelectasis of the underlying lung. The lesion was found to invade the right neural foramina at the D6-D8 vertebral level with compression and displacement of the spinal cord to the left side. It shows altered signal intensity appearing hyperintense on T2W1. Histopathological examination of tumor from the paraspinal mass shows features of a malignant round cell tumor with nuclear immunoreactivity of NKX 2.2. The patient was started on prephase therapy. Postprophase, he was referred to the nuclear medicine department for a positron emission tomography/computed tomography (PET/CT) scan for a work-up. PET/CT imaging was performed after an intravenous injection

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of 10 mCi ^{18}F-fluorodeoxyglucose (^{18}F-FDG). PET/CT revealed a metabolically active heterogeneously enhancing soft-tissue density mass lesion with areas of necrosis and calcifications in thoracopulmonary region involving the right posterior chest wall with the destruction of the right 6th– 8th ribs and adjacent vertebra with infiltration into the spinal canal abutting and compressing the cord. The maximum calculated standardized uptake value (SUVmax) of the lesion was 4.36 [Figure 1]. There was moderate right pleural effusion. Few metabolically active subcentimetric right extrapleural deposits and metabolically active bilateral supraclavicular, right paratracheal, subcarinal, and right retrocrural lymph nodes were also noted and deemed to be metastatic.

Discussion

PNETs can arise either from the central nervous system-PNET or from the periphery PNET.[4] Peripheral PNETs most commonly involve the chest wall pleura, pericardium, and soft tissue. PNETs of the chest wall were originally reported by Askin *et al*. in 1979 in 20 children;[1] since then, PNETs that occur within the thoracopulmonary region are named Askin's tumor. It is a rare malignant neoplasm, locally invasive in nature, and prone to destroying bone (ribs and scapula) and spreading to lymph nodes, adrenals, and liver. Askin's tumor infiltrating the spinal cord and presenting as a case of compressive myelopathy is rare. In a study of 104 patients with Askin's tumor by Laskar *et al*., asymptomatic swelling (43%) was the most common presenting symptom followed by pleural effusion. Lymphadenopathy was observed in 10.9% of cases and 25% of the patient presented with distant metastases. None of the cases presented with compressive myelopathy.[5] In another case series of 11 patients by Zhang *et al*., the most common clinical symptoms included fever, cough, chest pain, and suffocation.[6] In a review of the literature on peripheral PNET of the thoracic region, Anjankar reported 19 such cases with epidural mass in adults, with a female: male ratio of 1:1.7 and an average age of presentation at 30 years.[7] Presenting symptoms were back pain in all patients, followed by weakness in the limb (70%) and sensory loss (35%), and the diagnostic delay was approximately 4.5 months. There are not many case reports describing Askin's tumor ^{18}F-FDG PET/CT results. Xia *et al*. evaluated six patients of Askin's tumor with ^{18}F-FDG PET and observed that the lesion SUVmax varied from 4.0 to 18.6.[8] However, Kara Gedik *et al*. reported a SUVmax of 4.3 in their case report of a 73-year-old male with Askin's tumor.[9] In our case, the age at presentation was 24 years, and the patient reported signs and symptoms of compressive myelopathy which is a rare presentation in AT. ^{18}F-FDG PET/CT helped in the characterization of the metabolic activity of the tumor and in the detection of distant metastases. The metabolic activity of the tumor in our case was low (SUVmax 4.6) in concordance with Kara Gedik *et al*. Metastatic nodes were observed in the supraclavicular, mediastinal, and retrocrural regions. Supraclavicular and retrocrural metastatic nodes were also observed by Xia *et al*. in two of their six patients undergoing FDG PET scan. Winer-Muram *et al*. reported mediastinal lymph node metastases in two out of eight Askin's tumors,[10] whereas Sabaté *et al*. reported mediastinal lymph node metastases in one out of eight Askin's tumors.[11]

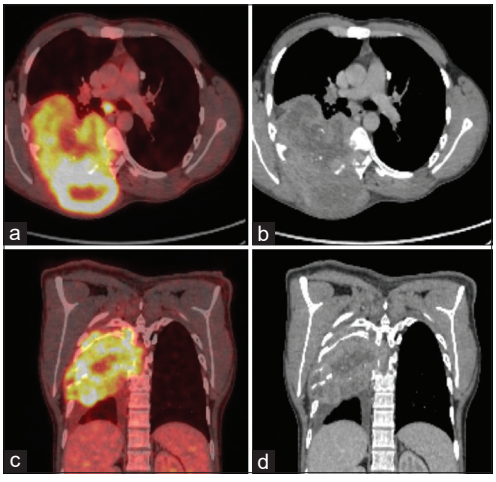

Figure 1: Axial fused positron emission tomography/computed tomography (a) and computed tomography (b) and coronal fused positron emission tomography/computed tomography (c) and computed tomography (d) images in a 24-year-old boy with lower back pain revealed an fluorodeoxyglucose avid heterogeneously enhancing thoracopulomary mass lesion in the right paravertebral region with areas of necrosis and calcifications infiltrating into the right thoracic cavity and into the right side of spinal canal abutting and compressing the cord. The adjacent ribs and thoracic vertebrae show destruction due to tumor infiltration

The Prognosis of AT is very poor. Because of rarity of the neoplasm, there are no consensus guideline for therapeutic management of the neoplasm, there is no consensus guideline. The treatment of Askin's tumor is multidisciplinary including surgery, radiotherapy, and chemotherapy aimed to control local disease and distant metastases.

Conclusion

This case emphasizes the fact that although Askin's tumor with compressive myelopathy is rare, it should be considered as one etiologic possibility in a young adult presenting with a case of paraplegia.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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