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CASE REPORT

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Posterior mediastinal Ewing sarcoma with multiple metastases on FDG PET/CT: a rare entity

Man Mohan Singh[1], Shashwat Verma[1*], Priyamedha Bose Thakur[1], Lavish Kakkar[1] and Satyawati Deswal[1]

Abstract

Background Ewing sarcoma and peripheral primary neuroendocrine tumors are aggressive neoplasms which consist of small, round, blue cells of neuroectodermal origin. They usually arise from the skeleton and consist of genetic mutations EWSR1 in chromosome 22 and FL1 gene on chromosome 11. Extraskeletal Ewing sarcomas (EES) are rare entities with most common sites of EES being extremities, head and neck region and retroperitoneum. Posterior mediastinal Ewing sarcoma is rare. For its evaluation, 18F-fluorodeoxyglucose positron emission tomography (18F FDG PET/CT) plays significant role in staging, management planning and prognostication.

Case presentation We describe a rare case of EES of posterior mediastinum in a 20-year-old boy who presented with signs of upper motor neuron lesion below D10 level. Contrast-enhanced magnetic resonance imaging (CEMRI) showed a heterogeneously enhancing posterior mediastinal mass in pre- and paravertebral region with intraspinal extension in D2-D4 levels. Fluorodeoxyglucose PET/CT showed a metabolically active mass occupying the superior, middle and posterior mediastinum on the left, displacing the trachea and esophagus toward the right side and causing complete collapse of the left lung. Posteriorly the mass was seen destroying the D2-D5 vertebrae with intraspinal extension at D2-D4 level. Metabolically active metastatic disease was seen in pleura, skull, D12 vertebra, right iliac bone and bilateral proximal femorae. Biopsy obtained from lung and adjacent pleura showed features of a round cell tumor positive for NKX 2.2, weak positive for FLI 1 and negative for PAN CK, LCA, Vimentin and TLE-1, suggestive of Ewing sarcoma. Based on these investigations, a diagnosis of EES of posterior mediastinum was made.

Conclusion Extraskeletal Ewing sarcoma of posterior mediastinum is a rare and aggressive entity. Management of metastatic EES comprises radiotherapy and systemic chemotherapy which reduces tumor burden and micrometastasis. However, response to treatment in metastatic EES is poorer than in localized disease with overall 5-year survival rates of less than 30%. Fluorodeoxyglucose PET/CT can be a useful tool to accurately detect the extent of local disease in the presence of atelectasic lung for radiotherapy planning as well as evaluating response to therapy.

Keywords Extraskeletal Ewing sarcoma, Mediastinum, FDG PET, Metastases, Pleural deposits

Background

Ewing sarcoma family of tumors are high-grade neuroendocrine tumors which usually arise from bony pelvis, femur and axial skeleton [1]. These tumors are known to have mutations in EWSR1 in chromosome 22 and FL1 gene on chromosome 11. Extraskeletal Ewing sarcomas are rare entities which comprise only 15% of all Ewing sarcomas, common sites being extremities, head and neck region and retroperitoneum. Even rarer

*Correspondence:
Shashwat Verma
drshashwat81@gmail.com
[1] Department of Nuclear Medicine, RMLIMS: Dr. Ram Manohar Lohia Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

among the EES are those of posterior mediastinum [2]. These are aggressive tumors with a high incidence of distant metastasis and recurrence after treatment [3]. We present a case of metastatic EES of posterior mediastinum in a 20-year-old boy presenting with sudden onset gradually progressive sensorimotor paraplegia. Posterior mediastinal Ewing sarcoma is a rare entity and not a common radiological differential diagnosis. For its evaluation, FDG PET/CT plays significant role in staging, optimal management planning, response evaluation and prognostication.

Case presentation

A 20-year-old male presented to Neurology department in December 2022, with complaints of pain in mid-back for one month and weakness of sudden onset in bilateral lower limbs for one week which progressed to a complete loss of function of both legs within another week. He also complained of incomplete voiding of urine and constipation for the past 2 days. There was no family history of Ewing sarcoma. Patient did not have any history of tobacco chewing or smoking. The genetic evaluation for Ewing sarcoma was not performed. On examination of the motor system, the patient had reduced power (1/5) along with increased tone in bilateral lower limbs and intact power (5/5) and normal tone in upper limbs. Sensory system examination showed absent sensory perception at D8 dermatome and below. Upper and lower superficial abdominal reflexes were absent bilaterally. Knee and ankle jerk were brisk (+++), and upper limb reflexes were normal (++). The blood parameters were within the normal limits except mildly elevated lactate dehydrogenase level. Contrast-enhanced MRI of the dorsolumbar spine was done, and images were reviewed by two radiologists having experience of more than 10 years. It showed a large ($12.4 \times 11.6 \times 8.1$ cm) heterogeneously enhancing mass in the posterior mediastinum invading the spinal canal in D2-D4 levels. Biopsy of the mediastinal mass and adjoining pleura showed tumor consisting of small, round cells with granular chromatin and scant cytoplasm. Immunohistochemistry showed NKX 2.2 positivity, FLI 1 weak positivity and PAN CK, LCA,

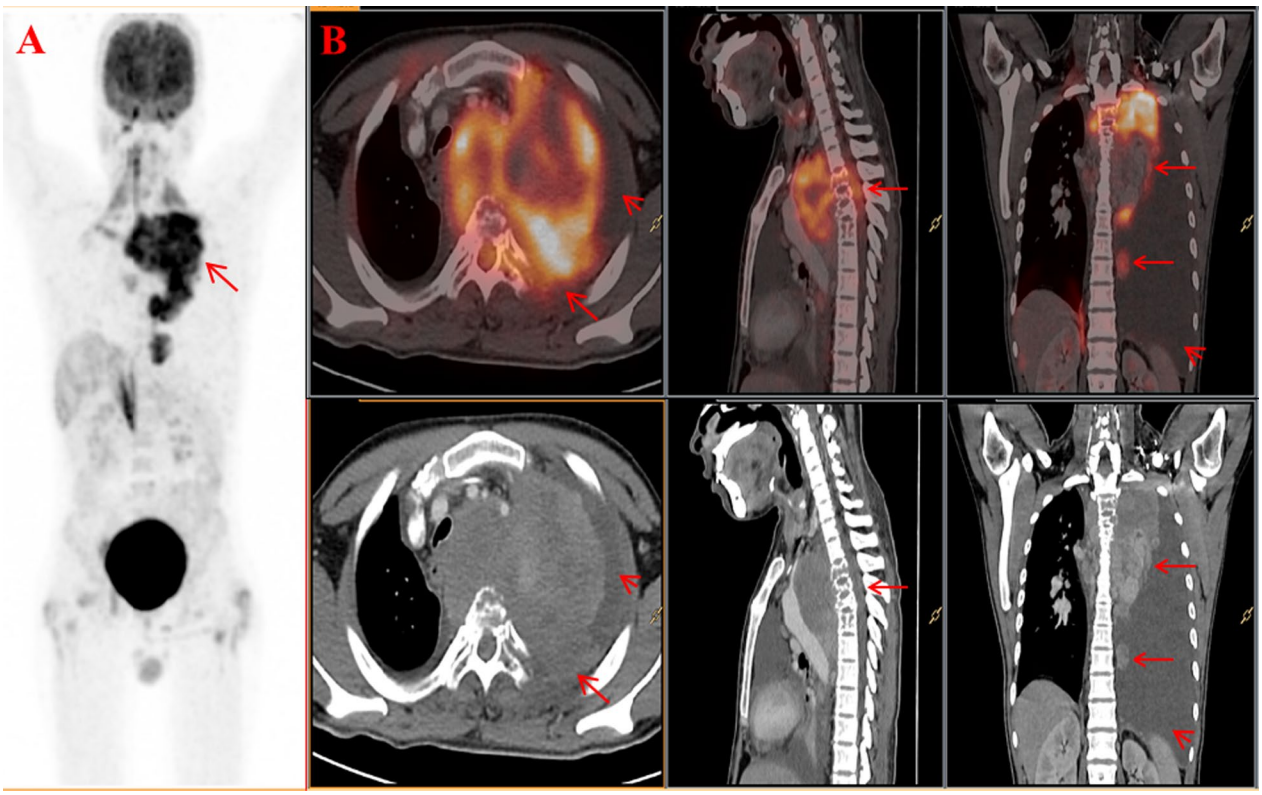

Fig. 1 A 20-year-old male, presented with pain in mid-back for one month, weakness of sudden onset in bilateral lower limbs for 1 week which progressed to a complete loss of function of both legs, incomplete voiding of urine and constipation for the past 2 days. Maximum intensity projection (MIP) image of 18F FDG PET scan **A** showing a large area of tracer uptake in the left hemithorax and midline region. Axial, sagittal and coronal PET/CT and corresponding CT images **B** showing metabolically active large posterior mediastinal mass with intracanalicular extension (in sagittal images) and left mediastinal pleural deposits (in coronal images) with left pleural effusion (arrow head)

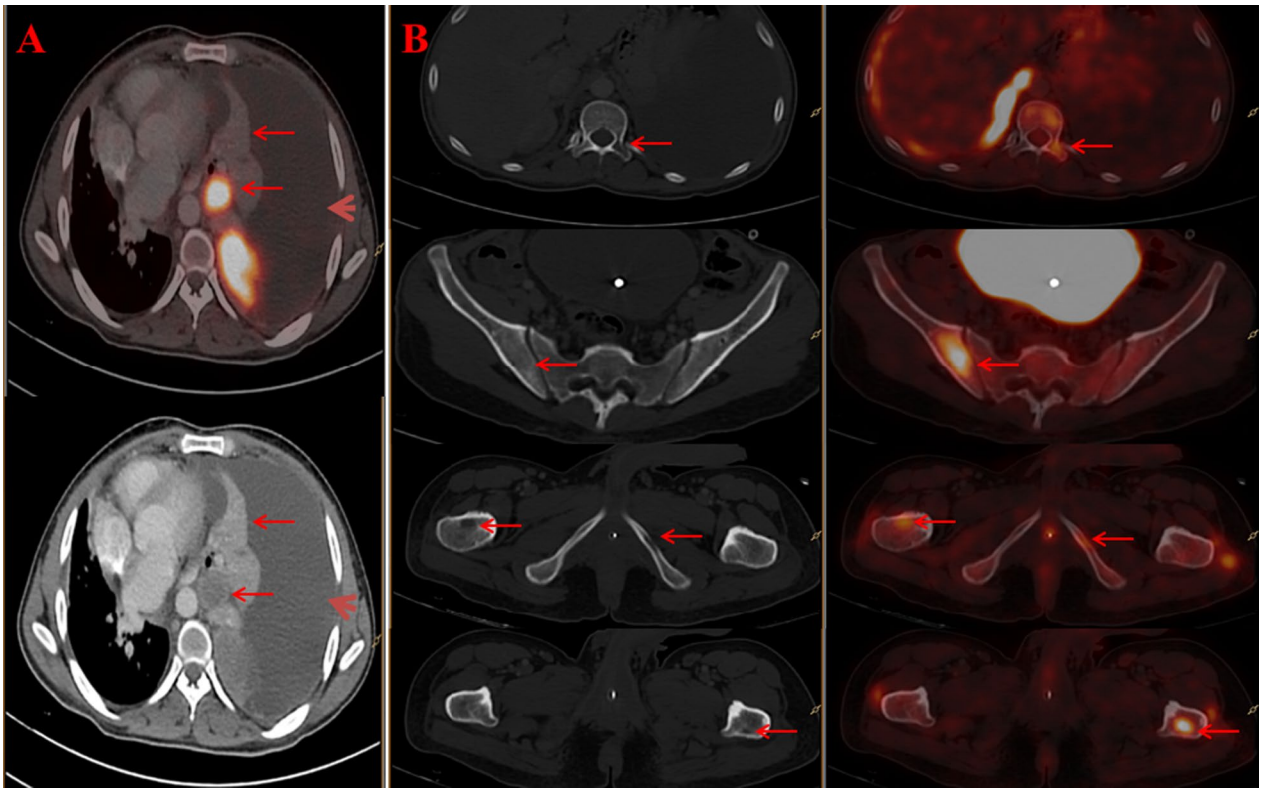

Fig. 2 PET/CT and corresponding CT images **A** showing metabolically active left hilar lymph node, left pleural effusion (arrow head) and left atelectatic lung. CT and corresponding PET/CT **B** showing FDG avid multiple lytic skeletal lesions (left transverse possess of D12 vertebra, right iliac bone, right femoral neck region, left ischiopubic ramus and left femoral neck region, respectively)

Vimentin and TLE-1 negativity, suggestive of Ewing sarcoma. Following this, the patient underwent FDG PET/CT and images were reviewed by the two independent nuclear medicine specialists having experience of more than 10 years. It showed a large FDG avid (SUVmax 15.2) heterogeneously enhancing mass occupying superior, middle and posterior mediastinum causing the trachea, esophagus as well as the heart to shift toward the right, abutting the arch of aorta and its branches and causing destruction of D2-D5 vertebrae posteriorly with intraspinal extension in D2-D4 levels (Fig. 1). There was a massive pleural effusion and collapse of the left lung. Metabolically active left pleural deposits (in Fig. 1B), hilar lymphadenopathy and multiple skeletal metastases were noted (Fig. 2). Minimal right-sided pleural effusion was also noted without significant FDG uptake. A diagnosis of Ewing sarcoma of the posterior mediastinum was made. Patient was then referred to medical oncology for further management; however the patient succumbed to the disease before treatment could be initiated. Age and gender were risk factors for Ewing sarcoma in this patient.

Discussion

Most common posterior mediastinal tumors are neurogenic in origin. Because of the anatomic location of posterior mediastinal tumors, these tumors can often reach a large size before becoming symptomatic. Mediastinal masses usually present with chest pain, breathlessness or cough. However, our patient presented with symptoms of extramedullary spinal cord compression which surfaced in a matter of few weeks. Neuroblastomas comprise most of these tumors causing intraspinal extension [4]. Ewing sarcomas of posterior mediastinum are rarely known to have intraspinal extension. It is not one of the initial differential diagnoses a clinician thinks of at the time of evaluation of a posterior mediastinal mass, as Ewing sarcoma appears radiologically indistinct from other common mediastinal masses such as soft tissue sarcoma or neuroblastomas [5]. Extraskeletal Ewing sarcoma has lung metastasis in the form of pleural deposits [6–8]. Lung metastasis in the form of parenchymal nodules or pleural deposits, which is more common, is not being well documented in EES because of paucity of literature. Pleural deposits may be more common. For accurately

differentiating the atelectatic lung from the tumor, PET/CT is beneficial, which may not otherwise be possible in CT. This advantage over conventional imaging can be important for planning radiotherapy, monitoring treatment response or assessing operability [9]. Extraskeletal Ewing sarcomas are aggressive tumors and are often unresectable or have distant metastasis at the time of diagnosis. Metastatic EES has a poor prognosis with a 5-year survival rate of less than 30%, and recurrent disease is always fatal [10].

Chest radiography may detect many pathologic conditions of posterior mediastinum. For further defining the image-specific features, the relationship to neighboring structures and providing differential radiological diagnosis, CT and MRI are the imaging modalities of choice [11, 12]. Fluorodeoxyglucose PET/CT is complementary to conventional anatomical imaging methods for the evaluation of mediastinal masses and also reduces unnecessary invasive investigations for diagnosis [13]. The PET/CT scan not only defines the primary lesion but also helps to detect local and distant disease involvement with high sensitivity and specificity. This case is staged accurately on FDG PET/CT and provides the optimal treatment planning. Fluorodeoxyglucose PET/CT has a proven role in the management of mediastinal tumors, helping to differentiate benign from malignant lesions when conventional imaging is inconclusive and for staging in the case of malignant mediastinal diseases [14].

Conclusions

Ewing sarcoma of posterior mediastinum is a rare diagnosis and is usually not included in the differential radiological diagnoses for a posterior mediastinal mass. These tumors have a late onset of symptoms and are often unresectable or have distant metastasis at the time of diagnosis. Fluorodeoxyglucose PET/CT helps in defining the primary lesion, local extent, regional and distant metastasis, i.e., staging for optimal treatment planning and restaging/treatment response assessment. Such patients should be investigated aggressively for providing them prompt treatment as the disease prognosis is poor.

Abbreviations

EES	Extraskeletal Ewing sarcoma
CEMRI	Contrast-enhanced magnetic resonance imaging
FDG	Fluorodeoxyglucose
PET	Positron emission tomography
CT	Computed tomography

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Not applicable.

Author contributions

All authors have read and approved the manuscript.

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Availability of data and materials

The dataset used and/or analyzed during the current study available from the corresponding author on the reasonable request.

Declarations

Ethics approval and consent to participate

Written consent to participate.

Consent for publication

Written consent for publication from study participant.

Competing interests

The authors declare that they have no competing interests.

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