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Metastatic Malignant Peripheral Nerve Sheath Tumor with Its Imaging Features

Saloni Rajkotia[1], Ritesh Ramesh Suthar[1,3], Abhishek Palsapure[1], Jahnavi Gandhi[2]

[1]Department of Nuclear Medicine, The Gujarat Cancer and Research Institute, Ahemdabad, India

[2]Department of Oncopathology, The Gujarat Cancer and Research Institute, Ahemdabad, India

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Abstract

Malignant peripheral nerve sheath tumors (MPNST) are rare and aggressive soft tissue sarcomas arising from the peripheral nerves. These tumors are associated with neurofibromatosis type 1 (NF1), and though they can occur sporadically, their diagnosis can be complex, especially when metastases are present. This case describes a young female patient with MPNST located in the dorsolumbar region and metastatic spread to the lungs, along with a history of acoustic neuroma.

Keywords: *FDG PETCT, MPNST, Nerve Sheath Tumor, Metastatic*

[3]Corresponding author: Ritesh Ramesh Suthar - received: 04.01.2025 - peer reviewed, accepted and published: 31.01.2025

Clinical Features

A 28-year-old female presented with a 6-month history of progressively worsening back pain localized to the dorsolumbar region. The pain was described as dull and constant, exacerbated by movement. The patient also experienced occasional numbness in her lower extremities. She had a past medical history of acoustic neuroma, diagnosed earlier. On physical examination, the patient had mild weakness in the lower limbs, along with tenderness over the dorsolumbar spine, but no palpable masses or neurological deficits were observed.

Radiological Findings

Initial radiographs of the dorsolumbar spine revealed an irregular mass in the paraspinal region, adjacent to the vertebral bodies. MRI of the spine demonstrated a large heterogeneous mass extending along the nerve roots, with areas of necrosis and contrast enhancement, suggestive of an aggressive lesion and lesion appeared to infiltrate surrounding tissues and involved the adjacent vertebrae.

Pathology Findings (Fig. 1)

On basis of MRI findings, biopsy of the dorsolumbar mass was done and it revealed histological features consistent with MPNST, including spindle-shaped cells arranged in fascicles, mitotic activity, and areas of necrosis. Immunohistochemical staining showed positive markers for S-100, vimentin, and nerve growth factor receptor (NGFR), confirming the diagnosis of MPNST. No evidence of NF1-associated mutations was found in this patient, suggesting a sporadic occurrence of MPNST despite the previous diagnosis of acoustic neuroma.

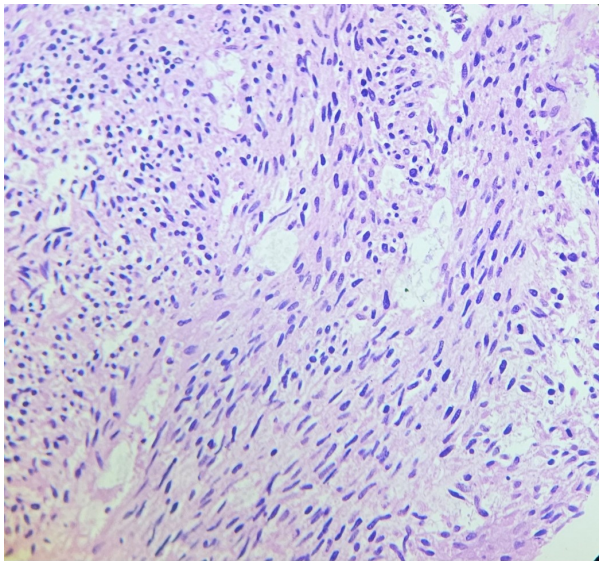

Fig. 1: H&E microscopic image shows tiny fragment with spindly cells in fascicles. Cells having moderate cytoplasm, oval hyperchromatic nuclei suggestive of malignant peripheral nerve sheath tumor.

FDG PETCT (Fig. 2)

A whole-body FDG PET/CT scan was performed to evaluate for metastasis. The scan revealed significant hypermetabolic activity in the primary dorsolumbar mass, consistent with malignancy. Additionally, multiple lung nodules were identified, showing intense FDG uptake, indicating metastatic disease. These findings were suggestive of lung metastasis originating from the primary MPNST.

Management and Intervention

Given the extensive local invasion and presence of lung metastases, the patient was considered for systemic chemotherapy, with a regimen including doxorubicin and ifosfamide. Additionally, palliative radiation therapy was recommended for the primary tumor site in the dorsolumbar region to alleviate pain and control local tumor growth. Surgical resection of the primary tumor was not feasible due to its size and infiltration into surrounding structures, and due to the metastatic disease.

The patient started chemotherapy and radiation therapy, with close monitoring for response. However, due to the aggressive nature of MPNST and the presence of distant metastases, prognosis remained poor, and the patient was counseled regarding the

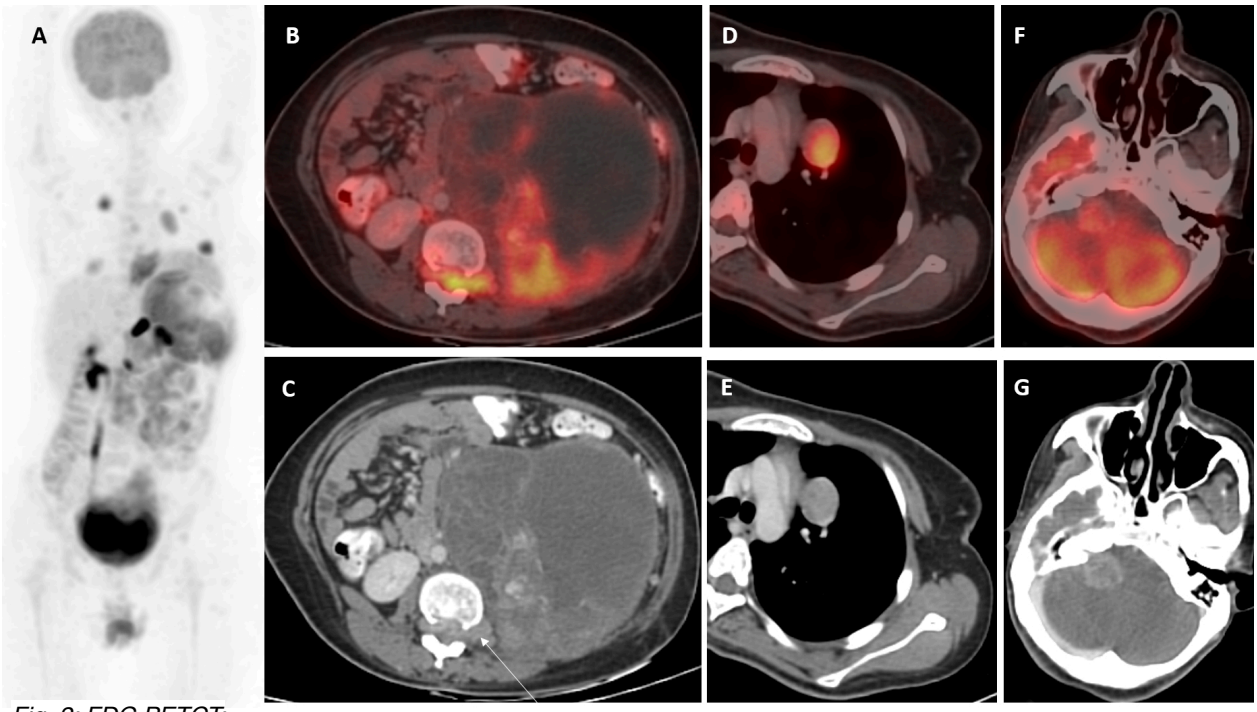

Fig. 2: FDG PETCT:
Image A: MIP image shows multiple focal FDG uptake in abdomen and thoracic region.
Image B&C: show a large FDG avid heterogeneously enhancing mixed solid cystic mass in left lumbar fossa of abdomen which showing connection with adjacent neural exit foramina (arrow in figure C).
Image D&E: show well defined FDG avid nodular lesion in lung suggestive of metastases.
Image F&G: show a FDG avid nodular lesion in right Cerebello-Pontine angle suggestive of acoustic neuroma.

likely outcome. Regular follow-ups were planned to assess treatment response and monitor for disease progression.

Discussion

MPNSTs are rare soft tissue sarcomas that arise from the peripheral nerves, often in the context of NF1, although this patient's case is sporadic. The radiological presentation, particularly on MRI and FDG PET/CT, is critical in assessing the extent of disease and potential metastasis (1). FDG PET/CT is especially useful in identifying distant metastases, as it can reveal early signs of lung involvement, which is common in MPNSTs. Pathologically, MPNSTs exhibit features such as spindle-shaped cells, mitotic figures, and S-100 positivity, confirming their neural origin (2).

Treatment for MPNST is challenging, particularly in cases with metastasis, as surgical resection is often not feasible. Chemotherapy and radiation therapy remain the mainstay of management (3), although the prognosis for metastatic MPNST is generally poor, with limited survival rates. Early detection and aggressive treatment are critical in managing these tumors, although the overall outcome in metastatic cases remains guarded.

Conclusion

This case highlights the complexity of diagnosing and managing MPNST, particularly when it presents with metastases. Radiological imaging, including MRI and FDG PET/CT, plays a crucial role in diagnosis, staging, and detecting metastases. Management typically involves a combination of chemotherapy, radiation, and palliative care, although prognosis remains poor in metastatic cases. Clinicians must be vigilant in considering the possibility of MPNST in patients with a history of neurofibromatosis or acoustic neuroma, especially when presenting with new or worsening symptoms.

Correspondence to
Dr Ritesh Ramesh Suthar
https://orcid.org/0000-0002-9383-069X
Dept. of Nuclear Medicine at The Gujarat Cancer & Research Institute
Civil Hospital Campus, Asarwa,
Ahmedabad-380 016.
Gujarat, INDIA
Phone :+91-79- 2268 8000
Fax : +91-79-2268 5490

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