Malignant Nerve Sheath Tumor with Retroperitoneal Extension

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ABSTRACT

Aim: To discuss a rare entity that was encountered along with its findings and outcome.

Background: Malignant peripheral nerve sheath tumors (MPNSTs) are sarcomas which originate from peripheral nerves or from cells associated with the nerve sheath, such as Schwann cells, perineural cells, or fibroblasts. These are rare but aggressive neoplasms. The MPNSTs are frequently seen in the head, neck, and upper extremities. Retroperitoneal cases are fairly rare and clinically difficult to be detected. These tumors have a very aggressive clinical course. The MPNSTs can arise de novo or from malignant transformation of benign nerve sheath tumors. The clinical course is usually short. In most cases, the diagnosis depends on the pathologic and immunohistochemical studies.

Case report: Here we report the case of a 42-year-old male patient who presented with swelling over lower back region and paresthesia over anterior aspect of left thigh since last 2 years, which aggravated over last 1 month. It was of pins-and-needle sensation type. Radiologically, the tumor was found extended from retroperitoneum into the spinal canal through the intervertebral foramina of D12–L1 and L1–L2 and compressing dura and its contents. Patient was treated surgically and histopathological examination confirmed MPNSTs.

Conclusion: The MPNSTs can arise de novo or from malignant transformation of benign nerve sheath tumors. The clinical course is usually short. In most cases, the diagnosis depends on the pathologic and immunohistochemical studies.

Keywords: Neoplasia, Retroperitoneal malignant peripheral nerve sheath tumor, Sensory impairment.


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INTRODUCTION

The commonly seen benign peripheral nerve sheath tumors are schwannomas and neurofibromas (NFs). Malignant peripheral nerve sheath tumors (MPNSTs) may arise de novo or from malignant transformation of benign counterparts. The MPNSTs represent 5% of all sarcomas, usually associated with NF in approximately 70% of cases. An association with previous irradiation has been observed. Diagnosis in these cases is difficult due to the admixture of benign and malignant histological components.

Nerve sheath tumors are common intradural extra-medullary lesions. Arising from nerve sheath, these tumors are benign in nature. Most of these are benign lesions. The MPNSTs are very rare but aggressive neoplasms. Most of these lesions are associated with neurofibromatosis where they may result from malignant degradation of plexiform NF. Incidence of MPNST ranges from 2 to 13%. About 5 to 10% of MPNSTs are soft-tissue sarcomas. Head, neck, and upper extremities are the most common sites involved in MPNST. Malignant nerve sheath tumors being highly aggressive usually present with very short history of back pain and neurological deficits. We present a 42-year-old male patient diagnosed with MPNST.
hip flexion and grade IV power in right hip flexion. There was sensory impairment for touch and pain in left L1, L2, L3 dermatomes. Knee jerk was impaired on both sides. On local examination, left paraspinal bulge was noted at L1–L2 level, which measured approximately 6 × 5 cm in size with ill-defined margins and was firm in consistency. Radiological examination was carried out. Computed tomography (CT) spine was suggestive of bony erosion in L2 vertebral body, left pedicle, facet, lamina, and spinous process along with posteroinferior portion of L1 vertebral body (Fig. 1). Magnetic resonance imaging (MRI) spine exhibited a lesion hypointense on T1-weighted and hyperintense on T2-weighted image, eroding L2 and a part of L1 vertebral body compressing cord at lower end of conus and cauda equine roots with paraspinal extension (Fig. 2). The lesion was extending from retroperitoneum via the spinal canal through the intervertebral foramina of D12–L1 and L1–L2 and compressing dura and its contents, invading L1 and L2 laminae, pedicles, facet joints, L1 and L2 vertebral bodies on the left side. No intradural extension was visible. Right and left L1, L2, and left L3 roots were encased by the tumor, infiltrating the paraspinal muscles and displacing kidney and other retroperitoneal structures. Tumor was firm in consistency, being highly vascular also, partially suckable, and causable. Posterolateral retroperitoneal part was clearly defined by a capsule. Near-total excision of tumor was done with the

Fig. 1: Computed tomography lumbosacral spine shows bony erosion of L2 vertebral body, left pedicle, facet, lamina and spinous process, and posteroinferior portion of L1 vertebral body

Fig. 2: Hypointense lesion on T1 MRI eroding L2 vertebral body and part of L1 body

Fig. 3: Hyperintensity involving vertebral body in T2 MRI sequence, suggestive of bony invasion by tumor

Fig. 4: In the retroperitoneum, kidney is pushed upward and laterally. There is a definite fat plane between the lesion and the kidney
help of urologist preserving renal pelvis system. Tumor infiltrating L1 and L2 body was excised and replaced by cage with bony chips. T12 and L3 transpedicular screw fixation was done (Fig. 5). There were no intraoperative or postoperative complications. Histopathologically, spindle cells were arranged in bundles and fascicles with mitotic figures, nuclear pleomorphism, coarse clumped chromatin, and conspicuous nucleoli (Figs 6 and 7). On conduction of immunohistochemistry, tumor cells were positive for S-100 (Fig. 8). These features were suggestive of malignant nerve sheath tumor. Based on the histopathology results, Ifosfamide and Adriamycin-based chemotherapy was started. Patient was under outpatient follow-up for 2 months.

DISCUSSION

Malignant peripheral nerve sheath tumors are sarcomas that originate from peripheral nerves or from cells associated with the nerve sheath, such as Schwann cells, perineural cells, or fibroblasts. A sarcoma is defined as a MPNST when one of the following criteria is detected:
- Arising from a peripheral nerve
- Derived from preexisting benign nerve sheath tumor (NF)
- Exhibition of Schwann cell differentiation on histological examination

Most arise in association with major nerve trunks, such as the brachial or sacral plexus or sciatic nerve. Approximately 50% arise in the trunk, 30% in extremities, and 20% in the head and neck; 50 to 70% patients suffering from NF encounter MPNST. Based on several studies in literature, the incidence of NF patients developing MPNST has been reported as from 2 to 29%. Incidence in the general population is around 0.001%. Up to 50% occur in patients with NF1, 10% are radiation induced, and 40% are sporadic. Our case appears to be unusual as there was no manifestation of neurofibromatosis-1. It is asymptomatic until later stage. The MPNSTs usually present as an enlarging palpable mass. Pain could be
variable. Rapid growth occurs most often in the presence of NF1 and raises concern for malignant degeneration of a NF. The MPNSTs arising from peripheral nerves may result in several types of clinical patterns, including radicular pain, paresthesia, and motor weakness. Large peripheral nerves, such as the sciatic nerve, the brachial plexus, and the sacral plexus are commonly associated with MPNSTs. Imaging modality of choice in these cases is MRI. Large tumors (>5 cm), invasion of fat planes, heterogeneity, ill-defined margins, and edema surrounding the lesion are more suggestive of MPNSTs. The CT scan helps in identifying bony changes. Though several radiologic imaging methods are helpful for identifying some features of NF, histological examination and immunohistochemical staining provide definitive diagnosis. In most cases, the diagnosis depends on the pathologic and immunohistochemical studies. The general appearance of MPNSTs is one of dense cellular fascicles that alternate myxoid regions. Marbleized pattern, i.e., swirling arrangement of intermixed dense and myxoid areas, has been described. Cells may be spindle-shaped with irregular contours. Or, they may be rounded or fusiform in shape. Nuclear palisading is seen in <10% of cases and, that too, only focally. Malignancy is determined by invasion of surrounding tissues, vascular structures along with nuclear pleomorphism, necrosis, and mitotic activity. S-100 expression was exhibited in approximately 50 to 90% of MPNSTs, though the staining pattern has been both focal and limited to few cells. The choice of treatment is surgical resection. The goal of the operation is to achieve complete surgical excision of the tumor; due to a high risk of recurrence owing to incomplete resection, postoperative irradiation and chemotherapy are necessary; however, they are often used as adjuvant therapy even if the tumor is completely resected. The reported 5-year survival rate for patients without NF1 is as high as 50%, which drops to 10% for MPNST patients with NF1. The 5-year survival rate of malignant schwannoma is low, primarily due to poor response of the tumors to available treatments and metastasis to the lungs and other organs.

CONCLUSION

Malignant nerve sheath tumors are very rare but aggressive neoplasias. As MPNSTs can arise from numerous cell types, its appearance can vary from one case to another. This makes diagnosis and classification quite difficult. The mainstay of treatment is surgical resection.

REFERENCES