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Case Report

Magnetic Resonance Imaging in a Malignant Peripheral Nerve Sheath Tumor- A case report

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Abstract: Malignant peripheral nerve sheath tumor (MPNST) is an extremely rare malignancy in the general population, occurring more frequently in patients with Neurofibromatosis type 1 (NF1). We report imaging features in a 45 year old female patient with Neurofibromatosis 1 and MPNST in upper part of fibular region. Plain radiography and Magnetic Resonance Imaging (MRI) were performed for a correct therapeutic planning. These findings were correlated with histopathological diagnosis which showed Malignant Peripheral Nerve Sheath Tumor (MPNST) with extensive necrosis. The diagnosis of MPNST is extremely difficult due to the lack of conclusive immunohistochemistry or unique chromosomal anomaly, universal distinctive histopathology and clinical criteria. The clinical, radiological and histological presentation of MPNST is important in its diagnosis and this case report will help clinicians in the diagnosis of this rare clinical entity.
Keywords: Malignant Peripheral Nerve Sheath Tumor, NF1, MRI, Plain radiography.

INTRODUCTION

Malignant peripheral nerve sheath tumor (MPNST) is an extremely rare malignancy. It usually encountered in patients with Neurofibromatosis type 1 (NF1) with an incidence of 2–5%, while in the general population it has an incidence of 0.001% [1, 2]. It arises from the Schwann cells of peripheral nerves, and it rarely involves the cranial nerves [3–5]. A malignant PNST generally presents as a soft-tissue mass that involves the major nerves and presents with pain and neurologic symptoms. A secondary malignant PNST may arise after radiation treatment, with a latent period that exceeds 10 years [6, 7].

Here, we present one such case of MPNST with swelling and pain over upper fibular region in a known case of Neurofibromatosis-1.

CASE HISTORY

A 45-year-old female who was a known case of Type 1 neurofibromatosis (NF1), also known as *von Recklinghausen disease*, presented to the surgical OPD of this hospital with swelling over lateral aspect of right leg near upper end of fibula. This swelling was gradually increasing in size since last 2 years and was associated with pain in the region of swelling since last 3 months.

At the time of presentation, physical examination revealed several cafe´-au-lait spots with multiple cutaneous neurofibromas over the entire body.

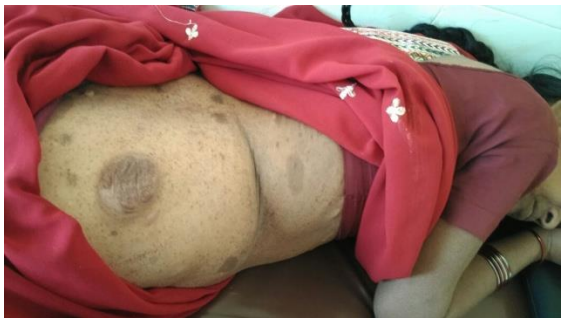

Image-1- Multiple café-au-lait spots with cutaneous neurofibromas were noted over the entire body.

She had a hematocrit of 24%, white blood cell count of 14.9 X 10^3/L, and albumin level of 1.6 g/dL. The remaining results of routine laboratory tests were unremarkable.

On local examination at surgical OPD, there was firm, tender swelling present on lateral aspect of right leg. So the patient was advised imaging investigations starting with the plain radiograph of the leg followed by MRI of the lesion.

Image-2: Cutaneous neurofibromas were present on the extremities also

MRI of right leg (Plain + Contrast) was done which showed large fairly well defined lobulated heterogenous intensity mass lesion along the lateral aspect of upper fibula1. It was hyperintense on T2WI and hypointense on T1WI. Lesion was extending posterior to fibula and tibio-fibular space. Mass was displacing adjacent muscle with mild edema around. It was scalloping and eroding fibular cortex with marrow edema around. Right peroneal nerve was not seen separately. Mass measured about 9.9 x 5.8 x 5.4cm and showed moderate heterogenous enhancement on post contrast study. These features were highly s/o Malignant Peripheral Nerve Sheath Tumor.

RADIOLOGICAL FINDINGS-
X-RAY

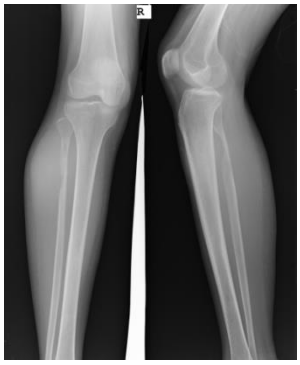

Image-3: Plain radiograph showed soft tissue swelling with cortical erosion of lateral aspect of fibula.

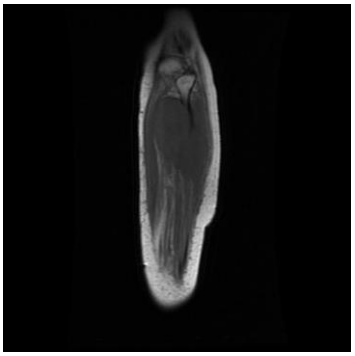

Image-4: T1 sag

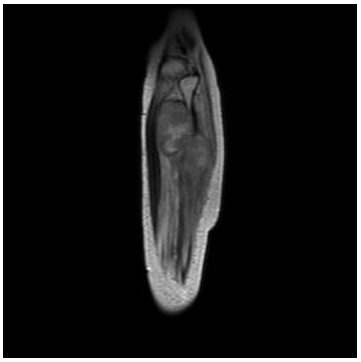

Image-5: T2 SAG.

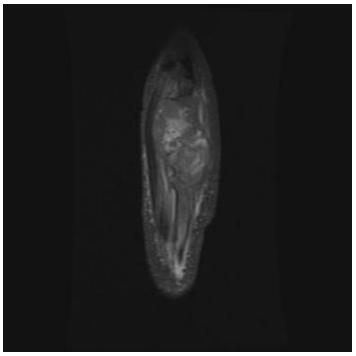

Image-6: STIR SAG

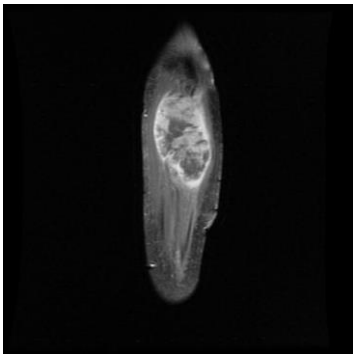

Image-7: T1 post contrast

HISTOPATHOLOGICAL EXAMINATION

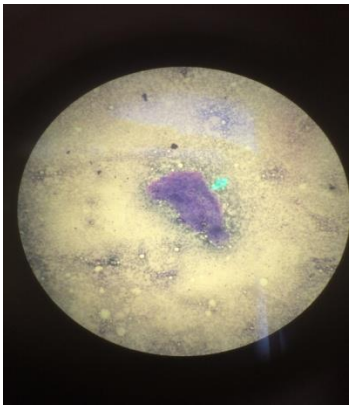

Image-8: Fragments of spindle shaped cells with elongated nuclei and coarse chromatin embedded in fibrillary matrix. Few cells are also singly scattered.

DISCUSSION

Malignant peripheral nerve sheath tumors (MPNST) have also been referred to as malignant schwannomas, neurogenic sarcomas, malignant neurilemmomas and neurofibrosarcomas. Malignant peripheral nerve sheath tumor is the current term used by the World Health Organization for this highly aggressive tumor. They most frequently affect patients who are 20–50 years old and represent 5–10% of soft-tissue sarcomas [8, 9]. A high proportion of malignant peripheral nerve sheath tumors (approximately 50%) occur in association with NF-1. On the other hand, only a small fraction of patients with NF 1 (approximately 5%) develop malignant peripheral nerve sheath tumors. [8, 9] These tumors generally involve the major nerve trunks and present with pain and neurologic symptoms, as well as a possible soft-tissue mass as it was in our case. Secondary malignant peripheral nerve sheath tumors can arise from prior radiation treatment, with a latency period of longer than 10 years [8, 9]. Primary intraosseous origin of malignant peripheral nerve sheath tumors is exceedingly rare, particularly involving a long bone; such lesions most commonly arise from the mandible [10, 11].

MPNST is a rare sarcomas arising from the Schwann cells of peripheral or exceptionally from the cranial nerves [3–7]. It can occur de novo or more frequently within pre-existing plexiform neurofibromas; thus patients with neurofibromatosis and plexiform neurofibromas warrant increased surveillance for development of these tumors [8].

Pathologically, malignant peripheral nerve sheath tumors are usually large fusiform masses, frequently with extensive central necrosis and hemorrhage. Although malignant and benign lesions cannot be reliably distinguished by imaging criteria, certain findings should raise the suspicion of a malignant tumor. Malignant peripheral nerve sheath tumors tend to be larger (>5 cm). They may exhibit ill-defined margins suggesting infiltration of adjacent tissues and associated edema [8] Heterogeneity with central necrosis on cross sectional imaging is common in malignant lesions although benign lesions with degeneration can also have a heterogeneous appearance. Similarly, calcification, more commonly associated with malignant lesions, can also be present in ancient schwannomas.

Malignant peripheral nerve sheath tumors can arise within a previous radiation field. Primary intraosseous malignant peripheral nerve sheath tumors show a lytic osseous lesion with cortical destruction and soft-tissue extension. T1-weighted MR images reveal low-to-intermediate signal intensity, and T2-weighted MR images show high signal intensity in the lesion. In general, radiologic findings are nonspecific; however, given an aggressive osseous lesion in the setting of neurofibromatosis, radiologists should consider an intraosseous malignant peripheral nerve sheath tumor [8, 9].

Some authors consider the fine needle aspiration biopsy useful diagnostically, but the definitive diagnosis is immunohistological. In particular, histology can allow differential diagnosis between MPNST and other types of fusicellular sarcoma on the basis of immunehistochemical markers, including S-100 protein and NSE [12, 13].

Malignant peripheral nerve sheath tumors are usually high-grade sarcomas and treatment typically involves a combination of surgical resection, adjuvant chemotherapy and radiotherapy. Local recurrence and metastatic disease are common complications. Our patient underwent a surgical excision of the tumor without any subsequent complication, followed by radiation therapy. Survival of MPNST is associated with complete tumor resection. The overall 5-year survival rate is about 44–50% which is affected by the patient's age, size of tumor, location of tumor, and margins affecting survival. Tumor size is the most reliable independent prognostic factor, with larger tumors having worse prognosis [14],

CONCLUSION

MPNST is a very rare disease and MRI is the technique of choice, because it presents a better contrast resolution than other modalities like CT scan. So, the knowledge and recognition of the characteristic signs for peripheral nerve sheath tumors can aid in the proper diagnosis and treatment of these lesions.

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