

Malignant Peripheral Nerve Sheath Tumour (MPNST) of Common Peroneal Nerve –Case Report

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Abstract: 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. World Health organization (WHO) coined the term MPNST replacing previous heterogeneous and often confusing terminology such as malignant schwannoma, malignant neurilemmoma and neurofibrosarcoma, for tumors of neurogenic origin and similar biological behavior. A combination of gross, histopathological and immunohistochemical studies are used for diagnosing these tumors. We herein present a case of MPNST arising from common peroneal nerve with neuro-vascular encasement and extensive soft tissue without bone involvement.

Keywords: Common Peroneal Nerve, Mpnst, Neurofibrosarcoma, Radical Surgery, Radiotherapy

1. Introduction

MPNST is a very rare tumor, with an incidence of 1 per 1,00,000 population and which constitutes between 3 to 10% of all soft tissue sarcomas[1]. The most significant contributions in understanding the clinical and pathological features of MPNST were studied by Mayo clinic investigators [2][3]. A combination of gross and microscopic findings along with immunohistochemical studies is commonly used to diagnose a case of MPNST [4]. Radical surgical resection is the treatment of choice in MPNST. A good three-dimensional clearance is mandatory for a successful outcome. Amputations are indicated only when wide excision is not feasible and in patients with severely compromised limb function.

2. Case Report

A 25 year old female presented to our department with mass over right lower limb with excruciating pain. Pain started first 6 months ago later swelling appeared and attained the present size. On physical examination swelling is of size 15*10cm, circumferential lesion involving anterior, lateral and posterior aspects of middle one third of right leg. On inspection swelling is looking like fungating mass with bleeding points with out skin covering, borders ill defined, appears to be arising from deep to deep fascia. On palpation there is tenderness, margins ill defined, swelling is immobile, movements of knee and ankle are painful. Routine investigations such as complete blood picture, urea, creatinine, LFT, serum calcium, phosphate were evaluated and are normal. X-ray shows that the tibia and fibula are not involved. Ultrasound of the lesion shows involvement of peroneal nerve and distal vascularity is compromised. MRI of the lesion cannot be done. Biopsy of the lesion was done and was diagnosed as MPNST.

Patient was given pre operative radiotherapy in order to shrink the size of tumor and plan for radical surgery.

Patient was advised amputation in view of large size and poor response to radiotherapy.



Figure 1: Clinical photo of patient with MPNST of common peroneal nerve





Figure 2: Clinical photos of the mass



Figure 3: X-ray right leg showing diffuse soft tissue shadow. Tibia and fibula appears to be normal

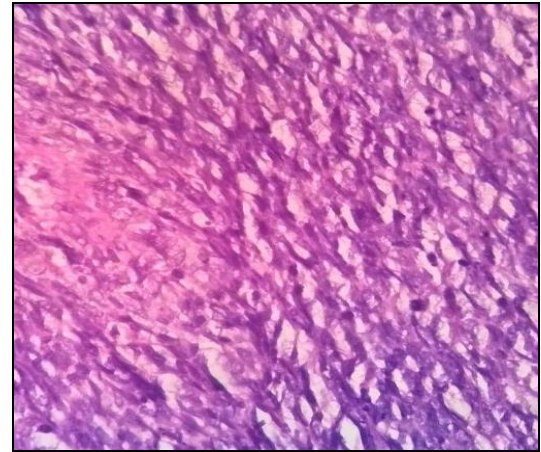
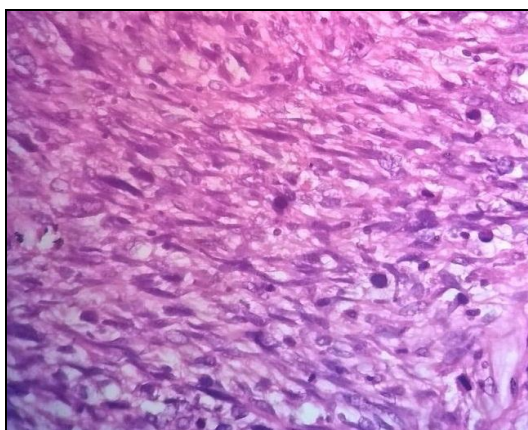
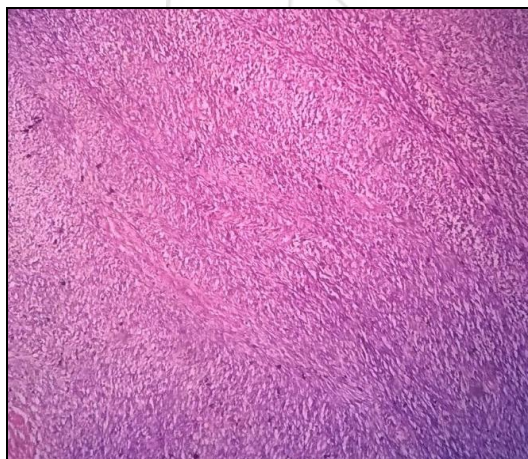


Figure 4: Multiple sections shows cellular lesion composed of sheets and fascicles of spindle cells. Cells containing mild pleomorphism. Few atypical mitosis are seen. Foci of hemorrhage and necrosis are seen. No evidence of lymphovascular invasion



Figure 5: Shows origin of the tumor from common peroneal nerve



3. Discussion

MPNSTs are rare soft tissue tumors that arise in proximity to large peripheral nerves and account for 3-10% of all soft tissue sarcomas [2,3]. The term MPNST was coined by the World Health organization (WHO) and is defined as any tumor that arises from a peripheral nerve, this term replaces previously used heterogeneous and often confusing terminology, such as malignant schwannoma, malignant neurilemmoma, and neurofibrosarcoma, for tumors of neurogenic origin and similar biological behavior[5]. These tumors arise from major or minor peripheral nerve branches or from the sheath of peripheral nerve fibers. Most of these tumors arise on the trunk, extremities or the head and neck region [4,5]. The pathologic diagnosis of MPNST is facilitated by features such as palisading arrangement, nuclear atypia, bizarre giant cells, mitotic figures and necrosis. These tumors have morphological heterogeneity and staining analysis of

such tumors reveals spindle cells with a fascicular pattern [6, 8]. Histological and immunohistochemical markers specific for MPNSTs are not available. The S100 protein is the antigen most commonly used to identify nerve sheath tumors of various types. However, S100 protein immunoreactivity is detected in only 50- 60% of MPNSTs and this protein is also expressed in a range of other tissues and tumor types [7]. Different markers are used to exclude other spindle cell tumors. Desmin and a-SMA are used to exclude smooth muscle tumors and CD34 and CD117 (c-kit) are used to exclude GIST [9].

A number of staging systems have been described. The most commonly employed staging system is the American Joint Committee on Cancer Staging System for Soft Tissue Sarcomas (see Table 1). Stage I essentially describes any low-grade small soft tissue sarcoma without evidence of metastasis. Stage II describes small high-grade tumors and large but superficial high-grade tumors without evidence of metastasis. Stage III describes high-grade large tumors which are deep. Stage IV includes any tumors with evidence of metastasis. One limitation of this staging system is that it does not reflect the tumor's anatomic location. This has been demonstrated to be relevant, especially in the setting of local recurrence [10].

Table 1: The American Joint Committee on Cancer (AJCC) Staging System for Soft Tissue Sarcoma, 6th Edition

Stage	Size	Depth	Grade	Metastases
I	Any	Any	Low	No
II	< 5cm, any depth OR > 5cm	Superficial	High	No
III	> 5cm	Deep	High	No
IV	Any	Any	Any	Yes

A biopsy is an integral part of the staging system. It offers both a histologic tissue diagnosis and the ability to determine the grade of the lesion. This information, in turn, permits adequate planning and adjuvant treatment such as radiation or chemotherapy. In addition, this information is incorporated into the tumor staging process which provides prognostic information with regard to the disease and treatment generalizations.

Radical surgical resection is the treatment of choice in MPNST. A good three-dimensional clearance is mandatory for a successful outcome. Amputations are indicated only when wide excision is not feasible and in patients with severely compromised limb function. Routine nodal dissection is not indicated. However, when a major nerve is identified, the cut end should be sent for frozen section to assess the tumor free margin of the resection. MPNSTs are generally considered chemotherapy and radiotherapy resistant tumors. However, there are reports of routine postoperative radiotherapy and even radiotherapy as a single modality alone for MPNST in literature [10]. In view of the rarity of this entity and conflicting reports, it is difficult to define the role of radiation in the management of MPNSTs. Currently, postoperative radiotherapy is recommended by as part of a uniform treatment policy for MPNSTs, much like other high grade soft tissue sarcomas[3,9], despite having clear surgical margins. Studies demonstrated 56%

disease free survival using combined surgery and radiation therapy for MPNST [11]. The indication of radiation treatment are biased towards patients having tumors with poor prognosis (high-grade or recurrent, deep seated and bigger size) and thus failing to show the statistically significant difference between radiated and non-radiated patients.

4. Consent

Patient is informed and explained about the procedure and written consent is taken for the procedure as well as publication.

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