

A Rare Case of Malignant Peripheral Nerve Sheath Tumour

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Abstract: Malignant peripheral nerve sheath tumor (MPNST) is a rare variety of soft tissue sarcoma of ectomesenchymal origin. World Health organization (WHO) coined the term MPNST replacing previous terminology, such as malignant schwannoma, malignant neurilemmoma, and neurofibrosarcoma. It has an incidence of 1 per 1,00,000 population and which constitutes between 3 to 10% of all soft tissue sarcomas. A solitary MPNST without neurofibromatosis is rarity and a MPNST arising from a small nerve ending is also rarity.

Keyword: Malignant peripheral nerve sheath tumor, neurofibrosarcoma, malignant schwannoma, malignant neurilemmoma, ectomesenchymal.

1. Introduction

Malignant peripheral nerve sheath tumor (MPNST) is a rare variety of soft tissue sarcoma of ectomesenchymal origin. MPNST occur with lifetime incidence of 8% to 13% in patients with neurofibromatosis type 1 (NF 1). MPNST rarely occur sporadically in general population. It typically affects in adult in the third to fifth decade of life. The lower extremities and retro peritoneum are most common sites, but MPNST can occur anywhere in body. They arise from a major or minor peripheral nerve branches or sheath of peripheral nerve fibres. Though these tumours have the highest rate of recurrence among soft tissue sarcomas, an adequate and a proper initial management improves the prognosis of the disease.

2. Case Report

A 50 year old man with presented to us with swelling in left thumb for 20 yrs to the outpatient unit. Swelling was noted in 1980, which was small in size and operated in same year. He was free from disease for 10 years, In 1990 swelling reappeared in same site. The swelling was insidious in onset and rapidly increased in size to attain the present

size. Swelling was rapidly growing, History of recurrent swelling in the left thumb No history of restricted movements in thumb, No history of pain, There was no familial history of neurofibromatosis, NF-1 (neurofibromatosis type I).

On Local Examination of Left Hand

A spherical shaped swelling situated in distal phalanx of left thumb, Size measures 4x4 cm, Swelling is firm, lobular in consistency, Margin is clearly defined, showing smooth surface, Skin over the swelling was normal with no sinuses or scars, Swelling was attached to deeper structure, Swelling was not warm, not tender, Skin over swelling not pinchable and not mobile.

FNAC OF SWELLING IN LEFT THUMB SHOWS Spindle shaped cells showing mild pleomorphism. FNAC of the swelling showed features of benign schwannoma

Surgery

Patient underwent a wide local excision of swelling during which it was found that the swelling was arising from a nerve twig. Swelling was infiltrating into surrounding area, distal phalanx was amputated leaving 1cm clearance.





Figure1: Swelling In Left Thumb

Histopathological Features

Malignant neoplasm composed of elongated neural cells with pleomorphic nuclei arranged in alternating areas of

highly cellular and sparsely cellular tumor cells with an admixture of collagen, numerous mitotic figures seen.

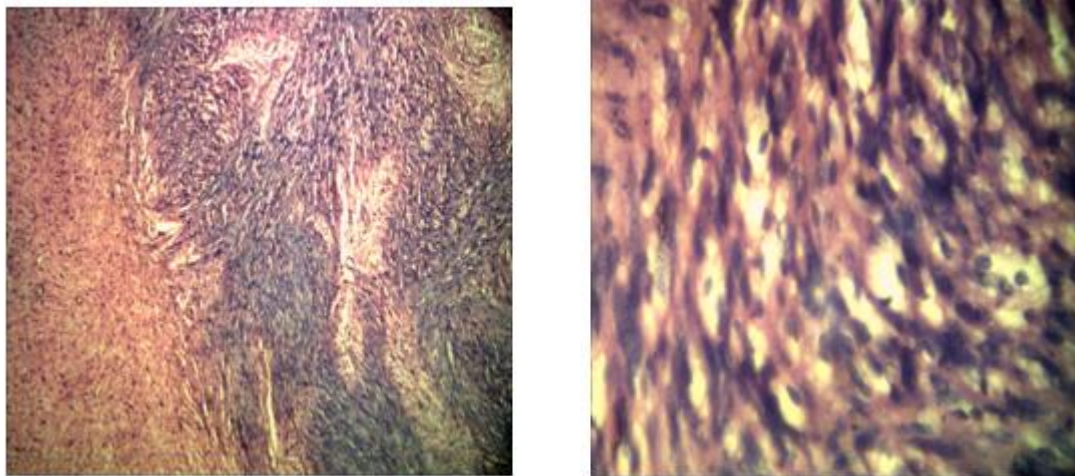


Figure 2: Histopathology of Malignant Peripheral Nerve Sheath Tumor Showing Elongated Neural Cells

Immunohistochemistry with S100, Proves the neural origin of the tumor

Post operative follow up

A Post-operative MRI showed no evidence of residual tumour or neurovascular infiltration. Patient was sent to a higher centre for chemo radiotherapy

histopathological, and immunohistochemical studies are used for diagnosing these tumors. Another interesting clinical feature of this tumor is multifocality and development of second primary tumors of same histology. Surgery is the main stay of treatment of this tumor though they are biologically aggressive in nature. In this article, we reviewed the case of a solitary MPNST without neurofibromatosis is a rarity and a MPNST arising from a small nerve ending is also a rarity.

3. Discussion

Malignant peripheral nerve sheath tumours are highly aggressive soft tissue sarcomas that rarely occur in the general population, but are much more common in patients with the hereditary tumour predisposition syndrome neurofibromatosis type 1 (NF1), which is caused by heterozygous mutations of the NF1 gene. NF1 patients have a lifetime incidence of MPNST of 8% to 13%.

These tumors often create diagnostic problems because of their cellular origin and histopathological similarities with other spindle cell sarcomas like monophasic synovial sarcoma, leiomyosarcoma and fibrosarcoma. They arise from a major or minor peripheral nerve branches or sheath of peripheral nerve fibers. These tumors may arise spontaneously in adult patients, although 5% to 42% of MPNST have an association with multiple neurofibromatosis Type-I. Thus, a combination of gross,

4. Conclusion

Malignant Peripheral Nerve Sheath tumors are aggressive tumors of neurogenic origin. It requires a combination of microscopic and immunohistochemical analysis for the diagnosis of MPNSTs. Surgery is the first line of management of MPNST and it is often important to achieve a clear margin in the initial surgery as it improves the prognosis.

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