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Original Research Paper

General Surgery

MALIGNANT PERIPHERAL NERVE SHEATH TUMOR IN A CASE OF MULTIPLE NEUROFIBROMATOSIS

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ABSTRACT Malignant peripheral nerve sheath tumors (MPNST) are rare, aggressive sarcomatous tumors that arise from peripheral nerve sheaths and show schwann cell differentiation (Thangadurai, 2017) [1]. They commonly arise among patients with neurofibromatosis I and following radiation therapy (Farid et al., 2014).

KEYWORDS : Malignant, Peripheral, Nerve, Sheath, Tumour, Case report

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNST) are classified as sarcomas that arise from peripheral nerves, a pre-existing peripheral nerve sheath tumour that has undergone differentiation or in concurrence with neuro-fibromatosis type I (NF I) syndrome [3]. NF 1 syndrome is characterized by the loss of the tumour suppressor gene, neurofibromin, and clinically the patient presents with multiple plexiform neurofibromas all over the body. The standard of care is an R0 resection with adjuvant radiotherapy.

MPNST has no gender predilection and commonly occurs between the 3rd and 6th decade of life [5]. Von Recklinghausen first described this condition in the late 1880s. It is characterized by the presence of café au lait spots (cutaneous hyperpigmentation) and multiple plexiform neurofibromas, which are slow growing peripheral nerve sheath tumours with a potential to rapidly increase in size, turn malignant and produce a mass effect. NF1 also presents with axillary freckling, optic gliomas, iris hamartomas termed Lisch nodules, bone dysplasia, cardiovascular abnormalities, and other malignancies (GIST, rhabdomyosarcoma, AML) [2]. The lifetime risk of a patient with neurofibromatosis I developing MPNST is 8%–13% [6]. A rapidly enlarging palpable mass in a previously asymptomatic patient with NF1 should raise the suspicion of malignancy. Pain, paraesthesia, weakness and other neurological deficits are common complaints the patient presents with. The most common site affected are the sciatic nerve roots. A size of more than 5 cm requires surgery because of its malignant potential and risk of metastasis to the lung. Pleural and bone metastasis is rare [2].

MRI is the imaging modality of choice. Tumours>5 cm, invasion of fat planes, heterogeneity, ill-defined margins and surrounding edema are highly suggestive of MPNST [7,8]. HRCT chest and a preoperative bone scan is required as a part of the metastatic work-up. FDG-PET works by assessing the intracellular glucose levels [9] in highly metabolic tumour cells.

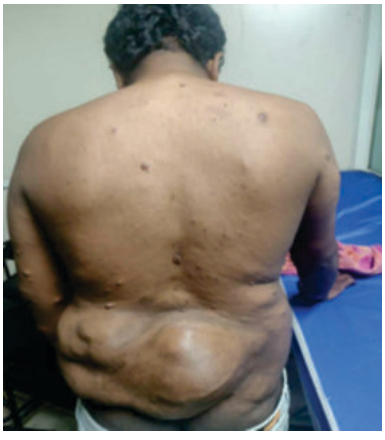
Fine needle aspiration and core needle biopsies play an important role in staging the disease. FNAC is preferred in cases where a recurrence is suspected.

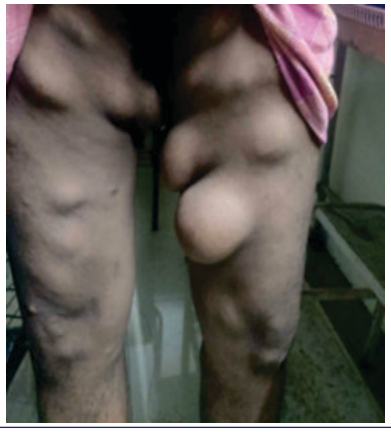
Case Study

A 27yr old male came with chief complaint of multiple swellings all over the body since 3yrs with largest swelling over lower back in the lumbar region which was gradually increasing in size since last 1year. Patient has no complaint of pain, fever, no recent loss of weight. Patient had a h/o surgery for multiple similar swellings in the past which again recurred. On examination largest swelling was of size cm over the lumbosacral region. Surface of the swelling was smooth, firm to hard in consistency not freely mobile. Skin over the swelling is normal and skin is pinchable. Cafe–lu-spots and axillary freckling is present. MRI revealed Large neurofibroma with sarcomatous degeneration(malignant peripheral nerve sheath tumour) with differential diagnosis as Liposarcoma. FNAC revealed the possibility of benign neurogenic neoplasm and advised biopsy for further diagnosis. Patient underwent excision of the multiple swellings including the largest swelling under General Anaesthesia.

Operative Findings

Elliptical incision is given over the lumbosacral region and incision deepened. A large tumour of size 15*8cm noted with high vascularity. Vessels are ligated and resection of tumour done and sent for histopathology which revealed GRADE 2(HIGH GRADE) PERIPHERAL NERVE SHEATH TUMOR with SPINDLE CELL SARCOMA. Rest of the other swellings were found to be lipomas on biopsy.





CONCLUSIONS

Complete surgical resection and achievement of tumour free margins is the mainstay of treatment. If the tumour size is more than 5cm, neoadjuvant radiotherapy is advocated to shrink the size of the tumour and reduce local recurrence [10].

Radiotherapy yields good results with respect to improved survival rates. Preoperative radiotherapy significantly reduces the size of the tumour. This approach reduces the total dose of radiation required and better tumour localization. Radiation causes tumour necrosis and reduces the chances of tumour spill . [11].

MPNST are difficult to manage because of their aggressive nature and the limitations in early diagnosis and management. Advances in computed tomographic scans and PET scans coupled with expert immunohistochemical analysis of lesions can accurately identify the stage of the disease and can predict its aggressive nature. Molecular targeted therapies following surgery for MPNST should be developed to render a patient disease free. In patients with Von Recklinghausens disease, malignancy must be suspected when a patient presents with the complaints as mentioned in this case report.

REFERENCES

1. Thangadurai Malignant peripheral nerve sheath tumor of chest wall. Univ. J. Surg. Surg. Spec. [Online] 2017;3(November (5)) [Google Scholar]
2. Fletcher C.D.M., Bridge J.A., Hogendoorn P.C.W., editors. WHO Classification of Tumours of Soft Tissue and Bone. IARC; Lyon, France: 2013. [Google Scholar]
3. Widemann B.C. Current status of sporadic and neurofibromatosis type 1-associated malignant peripheral nerve sheath tumors. Curr. Oncol. Rep. 2009;11:322–328. [PMC free article] [PubMed] [Google Scholar]
4. Farid M., Demicco E.G., Garcia R. Malignant peripheral nerve sheath tumors. Oncologist. 2014;19(2):193–201. [PMC free article] [PubMed] [Google Scholar] 7. Friedrich R.E., Kluwe L., Funsterer C., Mautner V.F. Malignant peripheral nerve sheath tumors (MPNST) in neurofibromatosis type 1 (NF1): diagnostic findings on magnetic resonance images and mutation analysis of the NF1. Gene. 2005;25(3A (May–June)):1699–1702. [PubMed] [Google Scholar]
5. Pilavaki M., Chourmouzi D., Kiziridou A., Skordalaki A., Zarampoukas T., Drevelengas A. Imaging of peripheral nerve sheath tumors with pathologic correlation: pictorial review. Eur. J. Radiol. 2004;52(3):229–239. [PubMed] [Google Scholar]
6. Hruban R.H., Shiu M.H., Senie R.T., Woodruff J.M. Malignant peripheral nerve sheath tumors of the buttock and lower extremity. A study of 43 cases. Cancer. 1990;66(6):1253–1265. [PubMed] [Google Scholar]
7. Dunn G.P., Spiliopoulos K., Plotkin S.R. Role of resection of malignant peripheral nerve sheath tumors in patients with neurofibromatosis type 1. J. Neurosurg. 2013;118:142–148. [PubMed] [Google Scholar]
8. Yang J.C., Chang A.E., Baker A.R., Sindelar W.F., Danforth D.N., Topalian S.L., DeLaney T., Glatstein E., Steinberg S.M., Merino M.J., Rosenberg S.A. Randomized prospective study of the benefit of adjuvant radiation therapy in the treatment of soft tissue sarcomas of the extremity. J. Clin. Oncol. 1998;16(1):197–203. [PubMed] [Google Scholar]