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Case report

A RARE CASE OF MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR

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ABSTRACT

Malignant Peripheral Nerve Sheath Tumours are tumours of ectomesenchymal origin often originating from major nerves or their nerve sheaths, they are commonly found in patients with neurofibromatosis-1 though sporadic cases have been reported. We report a rare sporadic case of MPNST in a 20 year old patient arising from the spinal accessory nerve.

Keywords: Peripheral nerve sheath tumour, malignant, neurofibromatosis.

INTRODUCTION

Malignant Peripheral Nerve Sheath Tumours are tumours of ectomesenchymal origin.[1, 2] MPNSTs, a term coined by the World Health Organisation represents collective tumours including malignant schwannoma, malignant neurilemmoma, neurofibrosarcoma and other neurogenic tumours that have the same biological behaviour.[3,4] They arise from major and minor nerves[5] or their nerve sheaths.[6, 7] As they are aggressive, surgery remains the primary line of management of MPNSTs.[8-10] They may arise as a sporadic variant or in patients with neurofibromatosis. The symptomology varies from a swelling to compressive symptoms and neurological deficits based on its size, location and tumour extension. We report a rare case of a sporadic Malignant Peripheral Nerve Sheath Tumour arising from a nerve twig of the spinal accessory nerve.

CASE REPORT

A 20 year old lady, with no history of NF-1, presented to the outpatient unit at Meenakshi Medical College Hospital and Research Institue, Kanchipuram with a rapidly growing non painful swelling in the left supraclavicular fossa with no history of pain radiating to her left arm (Fig. 1). The swelling was firm, lobular with well-defined margins and did not involve the skin. On putting the trapezius into contraction, the swelling became less prominent. A neck magnetic resonance imaging showed a well-defined intermuscular soft tissue intensity swelling with multiple axillary lymphadenopathies (Fig 2). FNAC of the swelling showed features of benignschwannoma and FNAC of the lymph nodes, done with ultrasound guidance showed granulomatous changes. Patient underwent a wide local excision during which it was found that the swelling was arising from a nerve twig supplying the trapezius (Fig.3). Histopathological examination showed pleomorphic spindle shaped cells arranged in intersecting fascicles with mitotic figures (Fig 4). Immunohistochemistry revealed positivity for S-100 and confirmed the diagnosis of MPNST with clear margins. A post-operative MRI showed no evidence of residual tumour or neurovascular infiltration.

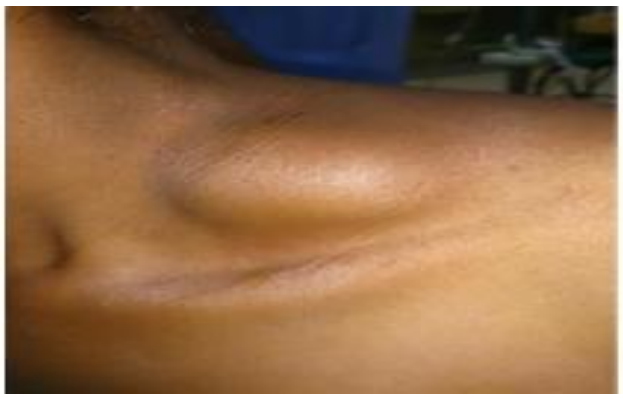

Fig1. Swelling in the left supraclavicular fossa

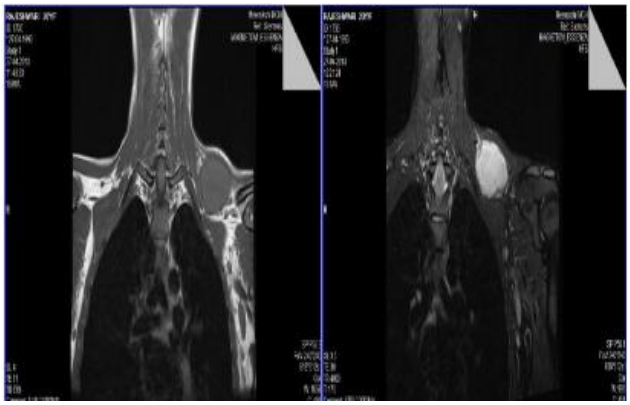

Fig 2: MRI of the left supraclavicular region showing a well defined lesion in the intermuscular plane

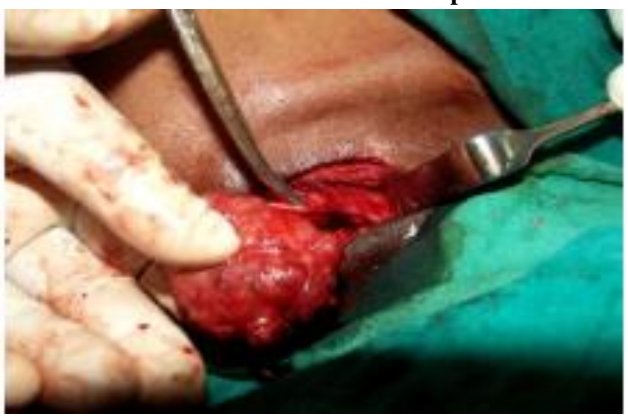

Fig 3: Tumour arising from the neural twig supplying the trapezius.

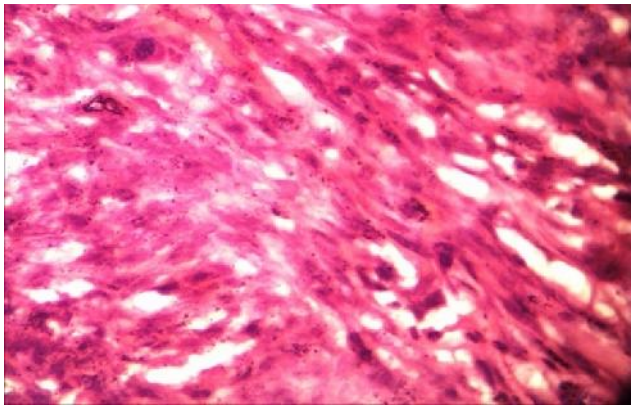

Fig 4: Showing spindle shaped cells with serpentine quality proving the neural origin with mitotic figures (40X)

DISCUSSION

MPNSTs of the head and neck are rare tumours and 70% of them arise in patients with von Recklinghausen disease, or neurofibromatosis.[1, 8, 11] It constitutes about one tenth of the soft tissue sarcomas and often considered a subgroup of the latter.[1, 2] Ducatman et al., [8] in his work declared that patients with von Recklinghausen disease had 4600 times risk of getting MPNST than those without VRHD. MPNSTs of the spinal accessory nerve are extremely rare. The diagnosis of MPNSTs usually requires a combination of microscopic and immunohistochemical studies.[8] Histologically, MPNSTs have a classic fascicular pattern of spindle cells displaying pleomorphism, mitotic figures and undifferentiation.[12] Immunohistochemical markers like vimentin, S-100 are used to confirm the diagnosis of MPNSTs[12]. Radical dissection with a clear margin is compulsory in the management of MPNSTs. The oncology consensus group recommends the use of post-operative radiotherapy despite achieving a clear margin. [10] Though these tumours have the highest rate of recurrence among soft tissue sarcomas, [13] an adequate and a proper initial management improves the prognosis of the disease. [14]

CONCLUSION

Malignant Peripheral Nerve Sheath tumours are aggressive tumours of neurogenic origin. It requires a combination of microscopic and immune histochemical 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. This case highlights the fact that a high suspicion of a sporadic MPNST should be kept in mind in dealing with patients with solitary cervical swellings.

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