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Case Report

Malignant Peripheral Nerve Sheath Tumour- Unveiling an Unknown Neck Mass

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Abstract

Malignant Peripheral Nerve Sheath Tumour (MPNST) is atypical and destructive sarcoma of the soft tissues which originates from peripheral nerves or their adjacent tissues. It is marked by invasiveness, malignancy, and sometimes linked to neurofibromatosis type 1 (NF1). Diagnostic complexity arises from varied histology resembling other sarcomas or benign nerve tumours. Treatment options are limited, especially for advanced cases, resulting in a grim prognosis. Surgical removal, often followed by radiation or chemotherapy, remains the primary localized treatment.

The patient in concern came with complaints of a progressively increasing swelling in the right side of neck for past 5 year, which was a solitary ovoid swelling extending from nape of neck to level of the right shoulder. CECT Neck revealed large exophytic mass involving right posterior triangle neck involving skin/ subcutaneous plane. Multiple site FNAC showed haemorrhagic aspirate with few stromal fragments and benign spindle cells. No diagnostic cells were seen. Total excision was done and sent for biopsy which revealed diverse histological features in multiple sections, some were tumour cells with serpentine shapes and myxoid backgrounds; pleomorphic hyperchromatic nuclei, including multinucleated giant cells displaying mitosis, some were round and pleomorphic tumour cells arranged in palisades or whorls. Metaplastic bone formation and necrotic areas were also present. Additionally, focal lymphocytic aggregations around blood vessels are observed. Patient was then sent for radiotherapy after evaluating for metastasis.

In conclusion, MPNST presents a formidable challenge with limited treatments and poor outcomes. Advancements in understanding its molecular basis and ongoing clinical studies offer hope for improved therapies. Collaborative diagnostic and management approaches are vital for enhancing patient care and results.

Keywords: Malignant Peripheral Nerve Sheath Tumour; Soft Tissue Sarcoma; Malignancy; Histopathologic Examination

Abbreviation

MPNST: Malignant Peripheral Nerve Sheath Tumour; HPE: Histopathologic Examination

Introduction:

Malignant Peripheral Nerve Sheath Tumour (MPNST) denotes malignancies emerging either from significant nerve branches or peripheral nerves, originating in Schwann cells or pleuripotent neural crest cells [1]. Constituting around five to ten percent of soft tissue sarcomas, these tumours contrast with common benign nerve sheath tumours in the head and neck (25-45%), as MPNSTs are infrequent in this locale [2]. Their prevalence is notable in areas like the parotid region and infratemporal fossa [3]. Occasional instances of superficial primary MPNSTs originating in cutaneous or subcutaneous layers exist. These superficial variants exhibit gradual growth over a prolonged period, followed by accelerated expansion [4].

Characteristically, MPNSTs present as palpable masses alongside alterations in the skin or symptoms localized to specific regions, such as vocal changes (in laryngeal primaries), difficulty swallowing (in oropharyngeal tumors), nosebleeds, nasal blockage,

or impairments in cranial nerve functions (in skull base tumors). Management entails the excision of the tumor through surgery, accompanied by supplementary radiotherapy, constituting the endorsed therapeutic strategy.[5] Despite surgery achieving clear margins, the recurrence risk remains high, necessitating comprehensive management strategies. The prognosis for advanced or metastatic MPNST cases is generally bleak due to limited treatment options. Surgical resection remains pivotal for localized tumours, often supplemented by adjuvant radiotherapy and/or chemotherapy. Although targeted therapies and immunotherapies are under exploration to enhance outcomes, further research is essential to validate their effectiveness.

Case Report

A 67-year-old male presented with a progressively enlarging swelling on the right side of the neck over the past five years. He had no history of trauma and reported no pain, restricted neck movement, breathlessness, voice alterations, swallowing difficulties, weight loss, or swellings elsewhere. On examination, a solitary ovoid swelling measuring 16x8 cm was observed, extending from the nape of the neck to the shoulder on the right side. The swelling exhibited variegated consistency, was non-tender, non-pulsatile, and mobile. The overlying skin appeared taut, stretched, and shiny, with prominent venous engorgement and a small ulcerative change near the tip (Figure 1,2). No cough impulse or fluctuation was noted. Auscultation yielded no significant findings.

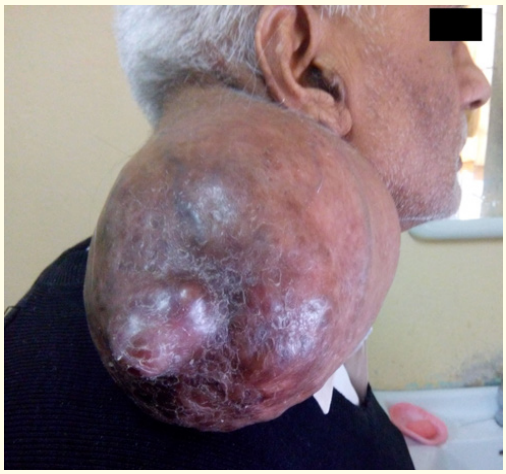

Figure 1: Shows a solitary ovoid swelling measuring, on the right side of the neck, with prominent venous engorgement and a small ulcerative change near the tip.

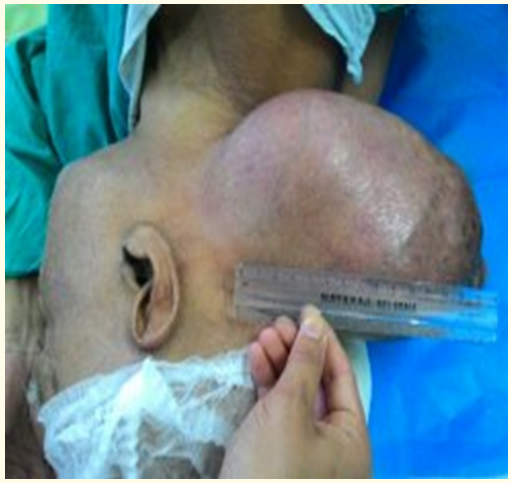

Figure 2: Shows solitary ovoid swelling measuring 16x8 cm; skin appeared taut, stretched ad vascular.

CECT Neck revealed a large exophytic mass (13 x 11 cm) involving the right posterior triangle of the neck and extending into the skin and subcutaneous plane (Figure 3). Multiple site Fine Needle Aspiration Cytology (FNAC) was performed, showing a haemorrhagic aspirate with fragments of stroma and benign spindle cells, devoid of diagnostic cells. As a result, the decision was made for a total excision and biopsy.

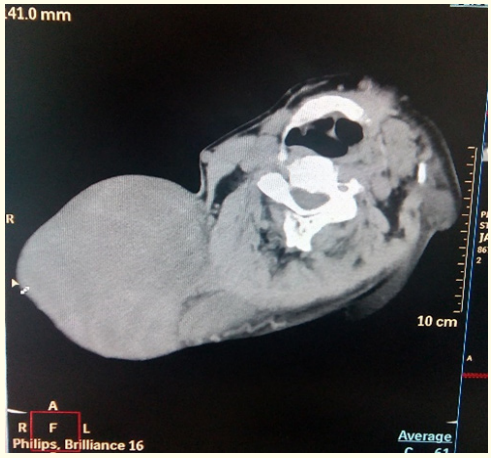

Figure 3: CECT Neck shows a large exophytic mass (13x11 cm) involving the right posterior triangle of the neck and extending into the skin and subcutaneous plane.

Under general anaesthesia, an elliptical incision was made around the base of the swelling, and the mass was dissected from the surrounding tissues (Figure 4,5). Feeding vessels around the base were ligated, and the mass was fully dissected and its pedicle ligated. Haemostasis was achieved, and excess skin was removed. A drain was placed, and the wound was closed in layers using sutures. The excised mass, weighing 2.1 kg and measuring 17 cm x 12 cm (Figure 6), was sent for histopathological examination (HPE). The postoperative period was uneventful.

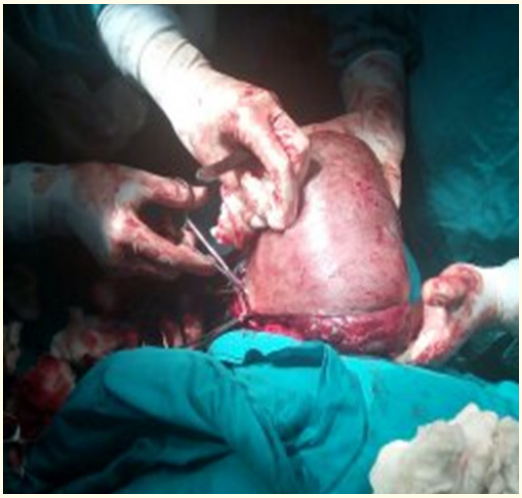

Figure 4: Shows operative elliptical incision made around the base of the swelling.

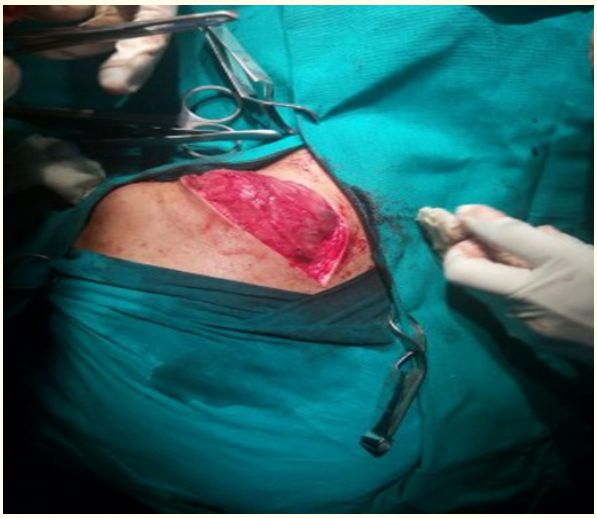

Figure 5: Shows operative dissection of mass from the surrounding tissues.

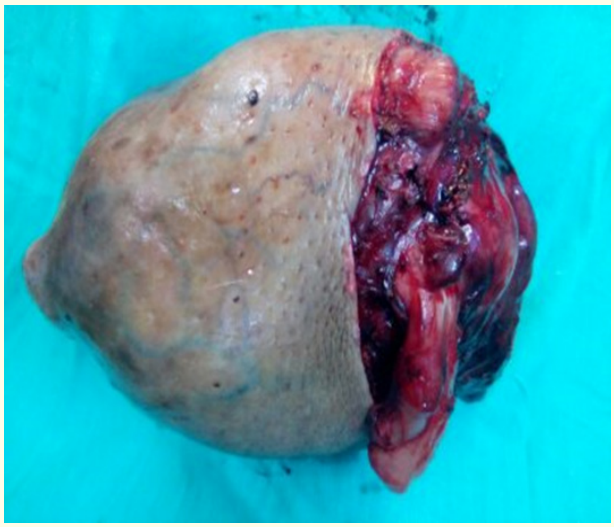

Figure 6: Shows the excised mass, weighing 2.1 kg and measuring 17 cm x 12 cm.

HPE revealed a divided mass, one part displaying necrotic grey-white changes, and the other part showing grey-yellow gelatinous material. No normal tissue was observed. Microscopic examination across multiple sections displayed varied histological features, including fascicular pattern and spindled shaped tumour cells amidst myxoid substance (Figure 7,8), with some cells exhibiting pleomorphic hyperchromatic nuclei and occasional multinucleated giant cells undergoing mitosis. Other regions showcased round pleomorphic tumour cells arranged in palisades or whorls, along with areas of metaplastic bone formation and necrosis. Focal lymphocytic collections were noted around blood vessels. Resected margins were tumour-free. The patient underwent a full-body CT scan for metastasis assessment, revealing no other metastatic sites. Subsequently, the patient was referred for radiotherapy at an advanced oncological centre. A one-year follow-up displayed no signs of recurrence.

Discussion

Malignant Peripheral Nerve Sheath Tumour (MPNST) is a relatively rare malignancy, constituting only about 5% of all malignant soft tissue tumors [5]. It stands out as a highly aggressive neoplasm that can either originate de novo from peripheral nerves or develop from preexisting benign neurofibromas [6]. Genetic mutations and alterations in key signalling pathways, such as the Ras pathway, are implicated in the pathogenesis of these tumours. In cases associ-

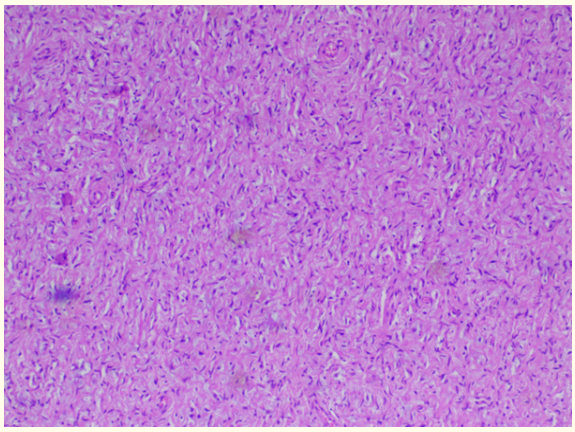

Figure 7: Shows histopathological examination at 100X: Tumour in fascicular pattern.

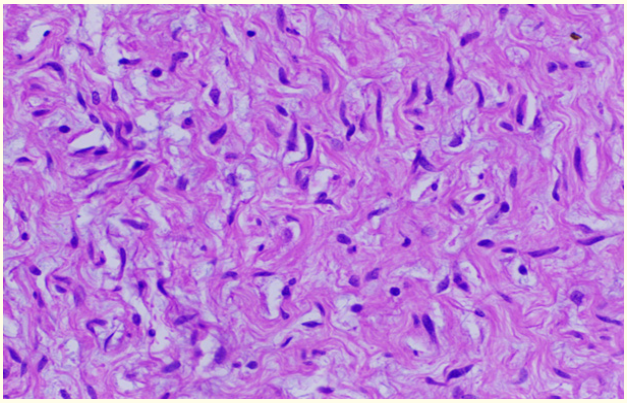

Figure 8: Shows histopathological examination at 400X: Spindled cells with wavy, buckled hyperchromatic nuclei.

ated with NF1, a mutation in the NF1 gene contributes to tumour development [7]. MPNSTs Can manifest in various anatomical regions, although they frequently arise in the extremities, trunk, or the head and neck area.

They present as palpable masses that may cause pain, neurological symptoms, or functional impairments. Diagnosis is challenging due to their varied appearance on imaging and histological examination.

The diagnosis of MPNST hinges on histological examination, further corroborated by positive responses to specific immunohistochemical markers [8]. This tumour type displays a broad histologic spectrum. They can commonly present as a densely cellular, spindle cell tumour exhibiting varying degrees of nuclear pleomorphism resembling benign neurofibromas to pleomorphic and hypercellular features indicative of malignancy. This variability contributes to diagnostic complexity. Immunophenotyping often reveals focal staining for S-100 and CD57 [9]. However, it's noteworthy that in our patient's case, the economic constraints hindered the performance of immunophenotyping. The prognosis is usually unfavourable, with a significant likelihood of reoccurrence and metastasis. The mainstay treatment for MPNST is surgical excision with wide margins, but complete resection can be difficult due to their invasive nature. Adjuvant radiotherapy and chemotherapy are often employed to lower the risk of local recurrence. The 5-year survival rate for MPNSTs is lower compared to other soft tissue sarcomas. In cases like ours, where economic factors limit the availability of comprehensive diagnostic tests, understanding prognostic indicators becomes all the more crucial. Indicators of poor prognosis include lesions exceeding 5 cm in diameter, deep-seated tumours, high-grade lesions, and instances of recurrence [9].

The alarmingly high recurrence rates of MPNSTs coupled with the limited efficacy of adjuvant therapies underscore the importance of a treatment strategy centred on en bloc resection with tumour-free margins. In this context, wide surgical excision emerges as the sole dependable treatment modality [10]. Given the propensity for both early and late recurrences, as well as the potential for second malignancies, postoperative radiotherapy (RT) becomes a necessity. It is an essential step in the comprehensive management of MPNSTs, addressing both immediate and potential future risks associated with this aggressive malignancy. Targeted therapies and immunotherapies are being investigated to improve treatment outcomes and extend survival.

Conclusion

MPNSTs are very rare but with poor prognosis hence advanced diagnostic tools are need of the hour for diagnosis especially in economically lower regions.

Declarations

Ethical statement: No unethical means were used to obtain the material. The patient was asked for permission to use his history and photos. The Head of Dept. of ENT, STNM has given permission on publication. The manuscript is not sent to any other journal.

- **Consent for Publication:** Consent for publication was taken from the patient and the Departmental head.
- **Availability of Data and Material:** Its available and all the material has been attached as figures.
- **Competing Interests:** None.
- **Funding:** None.
- **Authors' Contribution:** The first and second author worked on the collection of data, material and writing up of the article. The third author helped with the corrections on the material written.
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Bibliography

1. Gupta G., *et al*. "Malignant peripheral nerve sheath tumours". *Neurosurgery Clinics of North America* 19 (2008): 533-543.

2. Ho TP., *et al*. "Malignant peripheral nerve sheath tumour of the spinal accessory nerve". *The Journal of Laryngology and Otology* 113.10 (1999): 942-944.

3. Wong WW., *et al*. "Malignant peripheral nerve sheath tumor: analysis of treatment outcome". *International Journal of Radiation Oncology, Biology, Physics* 42 (1998): 351-360.

4. Allison KH., *et al*. "Superficial malignant peripheral nerve sheath tumor: a rare and challenging diagnosis". *American Journal of Clinical Pathology* 124.5 (2005): 685-692.

5. Ali SS and Ikram M. "Malignant Peripheral Nerve Sheath Tumor of Neck: A Case Report". *Research* 1 (2014): 1242.

6. Okada K., *et al*. "Clinical Relevance of Pathological Grades of Malignant Peripheral Nerve Sheath Tumor: A Multi-Institution TMTS Study of 56 Cases in Northern Japan". *Annals of Surgical Oncology* 14 (2007): 597-604.

7. Weber SM., *et al*. "R-Ras subfamily proteins elicit distinct physiologic effects and phosphoproteome alterations in neurofibromin-null MPNST cells". *Cell Communication and Signalling* 19.1 (2021): 95.

8. Cabibi D., *et al*. "Diagnostic and prognostic value of CD10 in peripheral nerve sheath tumors". *Anticancer Research* 29 (2009): 3149-3155.

9. Anghileri M., *et al*. "Malignant peripheral nerve sheath tumors: prognostic factors and survival in a series of patients treated at a single institution". *Cancer* 107 (2006): 1065-1074.

10. Minovi A., *et al*. "Malignant peripheral nerve sheath tumors of the head and neck: management of 10 cases and literature review". *Head Neck* 29 (2007): 439-445.