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Research Article

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Malignant Peripheral Nerve Sheath Tumour of the Scalp with Bone Metastasis

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ABSTRACT

Introduction: Malignant Peripheral Nerve Sheath Tumour (MPNST) is a malignant tumour arising from a peripheral nerve or showing a nerve sheath differentiation, with the exception of tumours originating from the epineurium or the peripheral nerve vasculature. Most of these tumours arise on the trunk, extremities, or head and neck regions. MPNST of the scalp is very rare.

Case Report: We, report the case of a 71-year-old woman with a lesion in the scalp over the occipital region with FNAC showing the features of MPNST. Wide local excision and split skin grafting was done. Histopathological analysis of the resected specimen strongly supported the diagnosis of MPNST. Patient received adjuvant radiotherapy and was disease free for 2 years and later presented with bone metastasis.

Discussion: MPNSTs are rare soft tissue tumours that arise in proximity to large peripheral nerves and account for 3-10% of all soft tissue sarcomas. Primary MPNST of the scalp is extremely rare and very few cases have been reported in literature. In this report, we have described the clinical and pathological characteristics of this rare tumor. We have also discussed the various treatment options offered.

Conclusion: Scalp MPNSTs are aggressive lesions, and multimodality approaches including surgery and adjuvant radiation are necessary to optimize outcomes. Every attempt should be made to obtain wide and negative margins, since positive margins are significantly associated with a poor outcome.

Key Words: Malignant peripheral nerve sheath tumour, Scalp, S100, Bone metastasis, MPNST, CD34

INTRODUCTION

Malignant peripheral nerve sheath tumours (MPNSTs) are uncommon malignant spindle cell tumours that account for 5% to 10% of all soft tissue sarcomas.[1,2,3] MPNST arise from nerve trunks located mainly in the trunk and extremities, such as the buttocks, thighs, brachial plexus, sciatic nerve and paraspinal region. Superficial primary MPNSTs with a cutaneous or subcutaneous origin represent a small subset of MPNSTs which is derived from cutaneous neurofibromas or small peripheral nerves.[1] Primary MPNST of the scalp is extremely rare, with only 14 cases reported to date in English literature.

Although the incidence of MPNST of the scalp is very low, it is highly malignant and is associated with a poor prognosis. We report a rare case of primary MPNST of the scalp and review the relevant literature regarding the clinical presentation, pathological features and outcome for MPNSTs in this anatomic location. In addition, we have also reviewed the various therapeutic strategies for treating these unusual lesions.

CASE REPORT

A 71-year-old woman presented with complaints of pain and increase in size of a swelling over the scalp in the occipital region of two months duration. She gave a history of a swelling over the scalp in the occipital region since childhood. Physical examination revealed a firm, mobile, non-tender mass. The haematological and biochemical tests were normal. FNAC from the lesion revealed discrete, as well as clusters of spindle cells exhibiting moderate degree of pleomorphism with the features of spindle cell sarcoma. Metastatic work up was negative. Patient underwent Wide Local Excision (WLE) of the lesion with split skin grafting.

Gross pathological examination of the WLE specimen revealed a whitish-grey lobulated glistening unencapsulated mass with deep resected margin 0.2cm away from the tu-

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mour. Microscopic examination revealed spindle cells exhibiting moderate degree of nuclear pleomorphism with high mitotic activity of 12 mitoses per 10 high-power fields (Fig I, II). Superior, inferior, medial and lateral soft tissue resected margins and skin were free of tumour. Immunohistochemical analysis of the tumour cells yielded positive staining results for S-100 and CD 34 and negative for Neuron Specific Enolase and Smooth Muscle Actin suggestive of a high grade MPNST, WHO grade IV.[7]

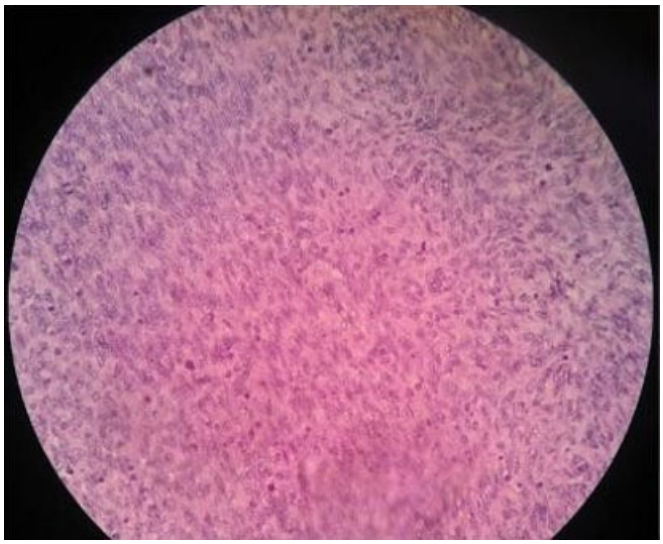

Figure I: Showing uniform spindle cells with hyper chromatic nuclei arranged in fascicles.

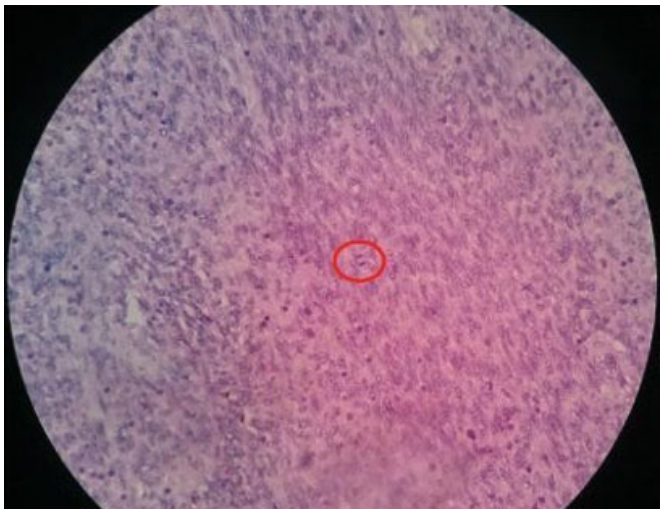

Figure II: Showing spindle cells exhibiting moderate degree of nuclear pleomorphism with high mitotic activity

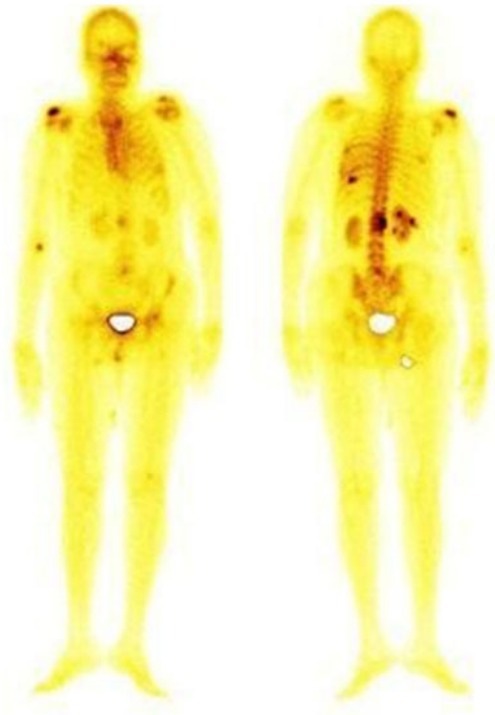

Figure III: Bone scan of the patient showing increased tracer uptake in right clavicle, multiple ribs, L1, L2, L5 vertebra, bilateral sacroiliac joints, upper half of shaft of right femur.

In view of the close deep resected margins and the high grade of the tumor patient was planned for adjuvant RT. She received 50Gy in 25 fractions at 200cGy per fraction. Patient was on regular follow- up. In May 2014, she presented with complaints of low back ache and increased frequency of urination. Bone scan showed increased tracer uptake in right clavicle, multiple ribs, L1, L2, L5 vertebra, bilateral sacroiliac joints, upper half of shaft of right femur (Fig III). Other metastatic work up was normal. She did not have any local recurrence. Patient was planned for palliative RT 30Gy in 10 fractions to lumbosacral spine. She completed palliative radiation to the spine without any complications. In view of advanced age and patient refusal, palliative chemotherapy was deferred.

DISCUSSION

MPNSTs are rare soft tissue tumours that arise in proximity to large peripheral nerves and account for 3-10% of all soft tissue sarcomas.[2,3] These tumours arise usually arise from major or minor peripheral nerve branches of the trunk, extremities, or the head and neck region. Primary MPNST of the scalp is extremely rare and less than 14 cases have been reported in literature. Superficial primary MPNSTs is derived from either a cutaneous neurofibroma or arise de-novo from small peripheral nerves.[1] Since our patient had a history of scalp swelling since childhood, the MPNST

probably arose as a result of malignant transformation of a pre-existing neurofibroma.

Palisading arrangement, nuclear atypia, bizarre giant cells, mitotic figures and necrosis facilitate the histologic diagnosis of MPNST. These tumours are characterised with morphological heterogeneity. Staining patterns of MPNSTs reveal spindle cells with fascicles. The S- 100, EMA, Vimentin (VIM) and CD34 antibodies are highly specific to MPNST.[5,6] For most MPNST cases, tumour cells exhibit differentiation towards Schwann cells, which is represented by immunoreactivity for S-100 protein and ultra- structurally by the presence of long cytoplasmic processes that are closely invested by a well-formed basal lamina. In some MPNST cases, tumour cells differentiate toward perineural cells, evidenced by immunoreactivity for EMA and ultra-structurally by the presence of tight junctions, abundant pinocytotic vesicles and an interrupted basal lamina. If tumour cells are not immunoreactive for either the S-100 protein or EMA, but are positive for vimentin, CD10, and CD34, these cells are considered to correspond to endoneurial fibroblasts. Desmin and SMA are used to exclude smooth muscle tumours.[8] Our patient showed positivity for both S 100 and CD 34 but SMA was negative ruling out a smooth muscle tumour.

There is a paucity of data regarding the management of MPNST of the scalp. The International Consensus Group has recommended the guidelines for the management of MPNST. The principles of management are similar to that of any other soft tissue tumours. Surgical excision of the lesion followed by reconstruction remains the mainstay of treatment. In case of MPNST of the scalp, reconstruction involves free flaps, skin grafts, or cranioplasty in case of significant calvarial destruction. The goal is to achieve complete surgical excision of the tumour with negative (wide) margins.[3] Our patient underwent wide local excision of the tumour with split skin grafting. In her case the deep resected margin was only 2mm. In a study by Kumar et al, positive tumour margins were the most important prognostic factor associated with a poor prognosis and every attempt should be made to obtain adequate negative margins of 2cms.[8] We offered the patient re-excision of the tumour but the patient refused further surgery. Adjuvant radiotherapy should be considered for all intermediate- and high-grade lesions as well as low-grade tumours with positive margins.[3] The supportive literature is generally in the context of sarcomas and not specific to MPNSTs. In view of the close deep resected margins and high-grade tumour, patient was planned for adjuvant RT.

In MPNST the role of chemotherapy is usually limited to the treatment of metastatic disease. The survival rates of patients with MPNSTs were significantly better for superficial tumours, such as MPNST of the scalp.[2,4] The possible reasons include the early detection and the greater possibility of achieving wide tumour margins without a disabling excision at these superficial sites. Moreover, clinical recurrences are also easier to detect because of the superficial location. However, the metastatic rates are similar to deep seated MPNSTs. Our patient developed bone metastasis 2 years later.

CONCLUSION

MPNSTs should be considered in the differential diagnosis for any patient with a rapidly enlarging and painful soft tissue mass of the scalp, particularly with a background of pre-existing neurofibroma. Scalp MPNSTs are aggressive lesions, and multimodality approaches including surgery and adjuvant radiation are necessary to optimize outcomes. Every attempt should be made to obtain wide and negative margins, since positive margins are significantly associated with a poor outcome.

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Conflict of interest

Nil

Consent

Written informed consent was obtained from the patient to publish this case report.

AUTHORS' CONTRIBUTION:

Abhishek Raghava K S has collected the case details, pathological images, bone scan images and conducted a comprehensively literature search and added to the intellectual content of the study.

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