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Case Study

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NERVE SHEATH MYXOMA AT AN UNUSUAL SITE- A CASE REPORT

Dr. Priyanka Anand[1]* and Dr. Poonam Sahni[2]

[1]Consultant Pathologist, Saral Diagnostic Centre, Pitampura, New Delhi.
[2]Director & Head of Department, Senior Consultant Pathologist, Saral Diagnostic Centre, Pitampura, New Delhi.

***Corresponding Author: Dr. Priyanka Anand**
Consultant Pathologist, Saral Diagnostic Centre, Pitampura, New Delhi.

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ABSTRACT
Nerve sheath myxoma is a rare slow growing benign tumor of peripheral nerve sheath origin, commonly involving extremities. It was first described by Harkin and Reed in 1969. It is usually seen in middle aged females. There is a lot of confusion regarding the line of origin and its relationship with Neurothekeoma. Immunohistochemistry plays an important role in distinguishing. Complete excision with a free tumor resection margin is the treatment of choice. We present a case report of Nerve sheath myxoma due to its rarity and an unusual location in the scalp.

KEY WORDS: Nerve sheath, scalp, neurothekeoma, myxoma, immunohistochemistry.

INTRODUCTION

Nerve sheath myxoma is a rare slow growing benign tumor of peripheral nerve sheath origin, commonly involving extremities with a predilection of fingers. It rarely occurs in the head, neck and intraspinal space.[1,2] It was first introduced by Harkin and Reed in 1969.[3] It has a female preponderance and is usually seen in middle aged.[4] We hereby report a case of Nerve Sheath Myxoma of scalp.

CASE PRESENTATION

A 48 year old female presented with a nodular lesion the scalp with a history since 3 years. The mass 1.5cm in diameter, was firm and mobile. Excision was done and the specimen was sent for histopathological examination. On gross examination, a skin covered soft tissue piece was received measuring 3x2x1.5cm and on cut section, a well circumscribed area, tan to pale yellow in color, homogeneous in appearance was noted measuring 1.5x1.5cm. Microscopic examination showed epidermis lined by stratified squamous epithelium. Underlying subcutaneous tissue showed a well circumscribed, focally encapsulated tumor arranged in lobules, nodules and fascicles with a geographical Tumor cells were spindle shaped, having round to ovoid, vesicular nuclei and abundant pale eosinophilic in a background of predominantly myxoid matrix. Cells were separated by prominent hyalinized collagen rich fibrous septa. No distinct perineural invasion was noted. No tumor necrosis was seen. No atypia was apparent. No mitotic figures were identified. On Immunohistochemistry, SOX-10 was immunoreactive in neoplastic cells (Score 3+). Also schwann cells in NSM were diffusely positive for S100 (Score 3+). Vimentin was also positive (Score 4+). Ki-67 was 4-5% in highest proliferation areas. CD10 was negative. In view of these findings, a diagnosis of Nerve sheath Myxoma (NSM) was rendered.

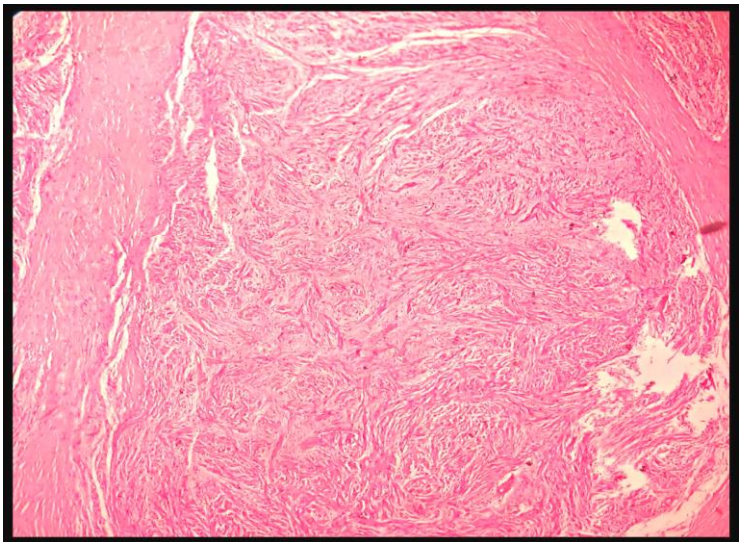

Figure 1: Microphotograph showing a well demarcated lesion comprising of nodules and lobules. (H & E Stain, 4x).

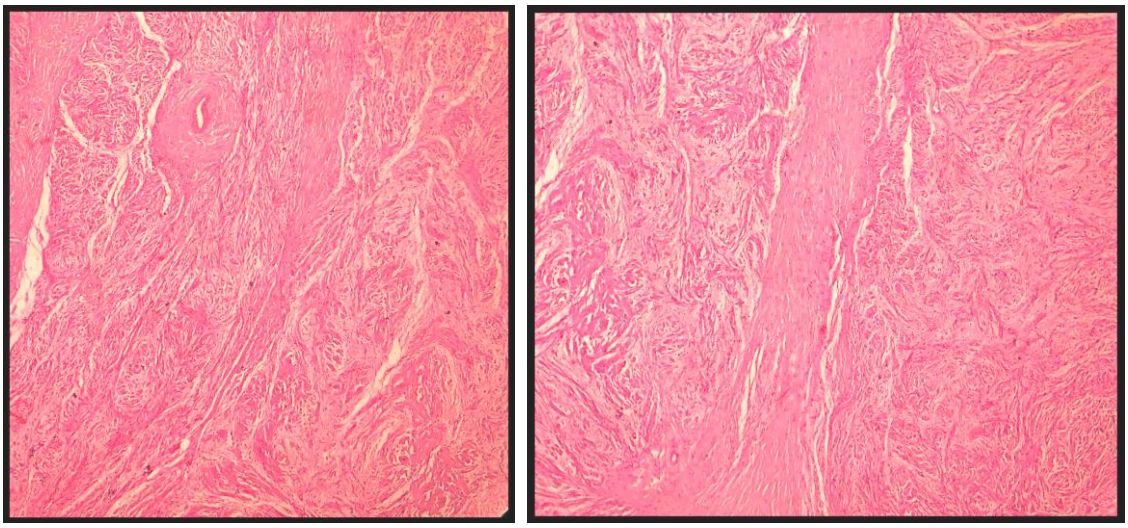

Figure 2: Microphotograph showing nodules separated by prominent fibrous septae. (H & E Stain, 4x and 10x).

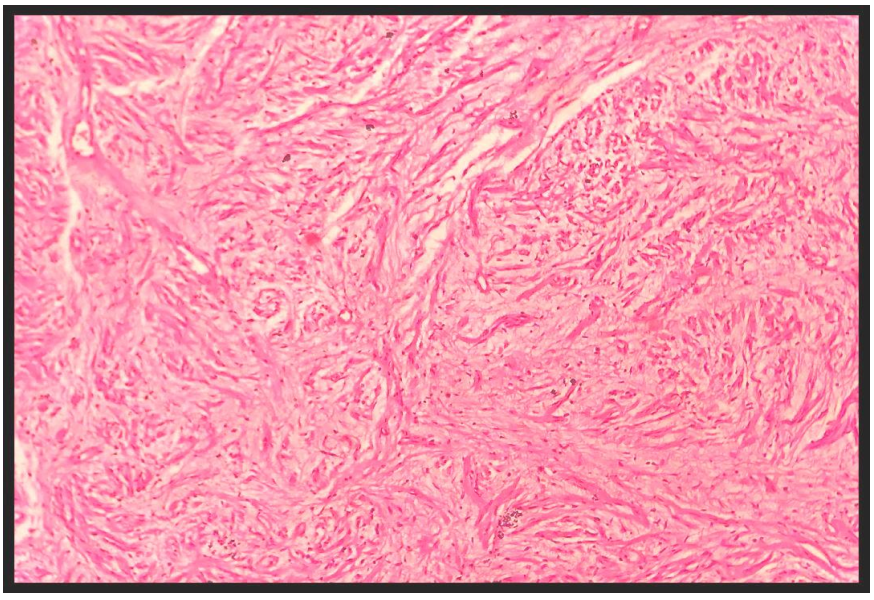

Figure 3: Microphotograph shows myxoid matrix in the background. (H & E Stain, 10x).

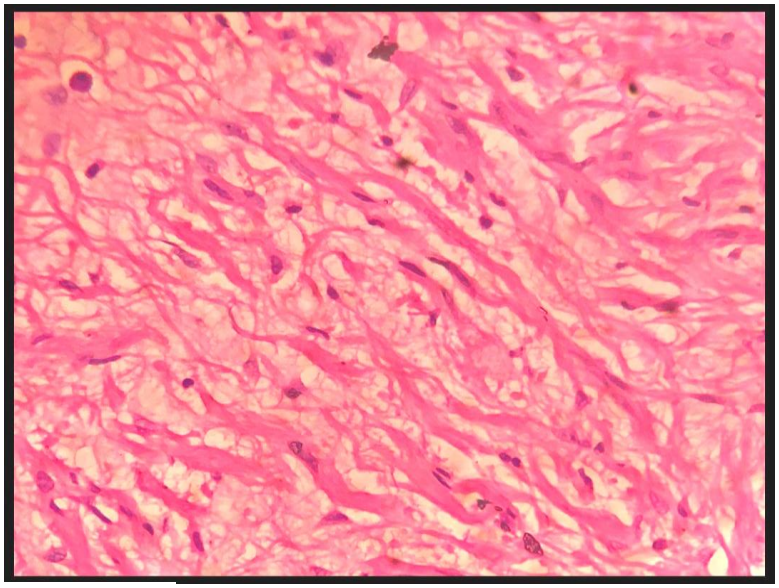

Figure 4: Microphotograph showing spindle shaped cells, having round to ovoid, vesicular nuclei and abundant pale eosinophilic cytoplasm. (H & E Stain, 40x).

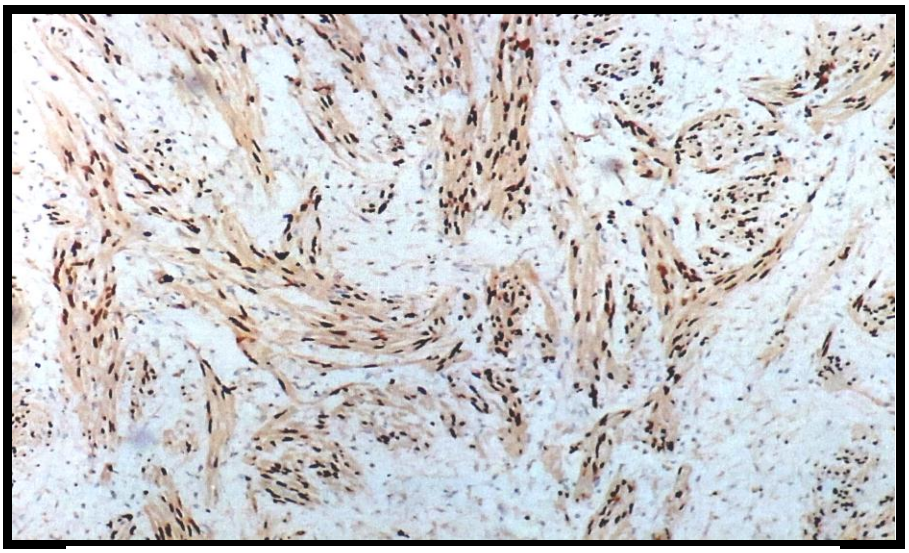

Figure 5: Tumor cells showing positivity for SOX-10. (DAB, 10x).

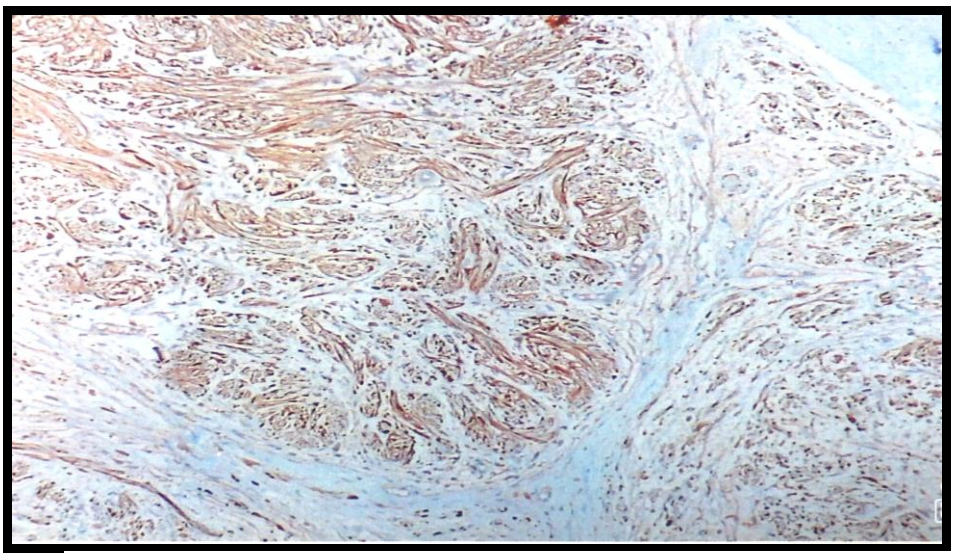

Figure 6: Microphotograph showing tumor cells positive for S100, Score 3+. (DAB, 10x).

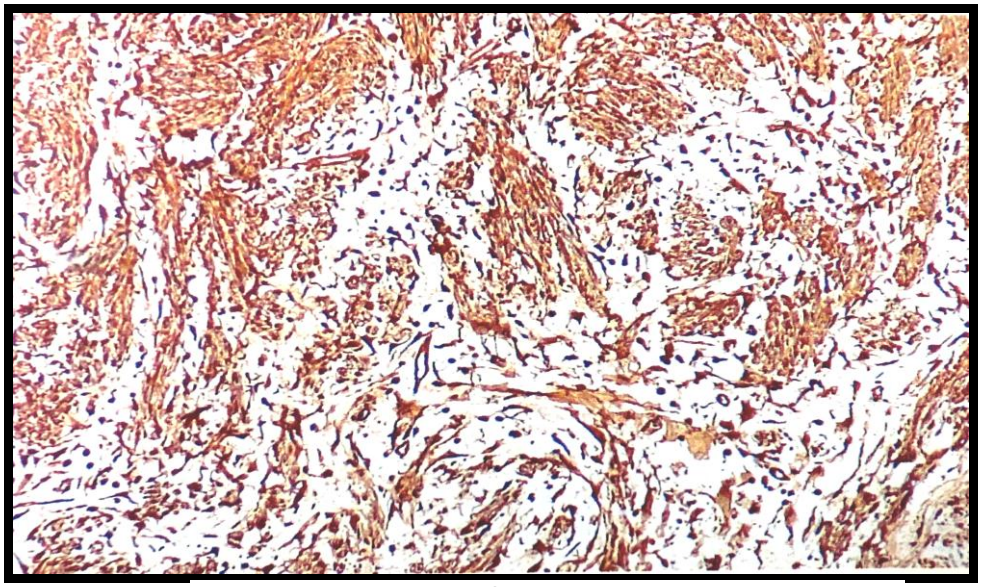

Figure 7: Tumor cells are positive for Vimentin, Score 4+. (DAB, 10x).

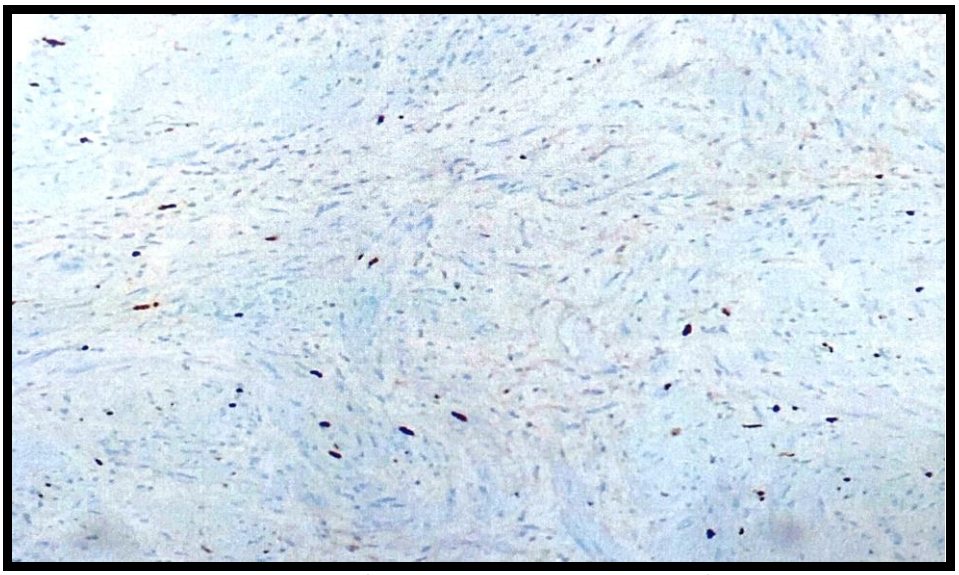

Figure 8: Tumor cells showing 4-5% proliferation rate in highest proliferation areas. (Ki- 67, DAB, 10x).

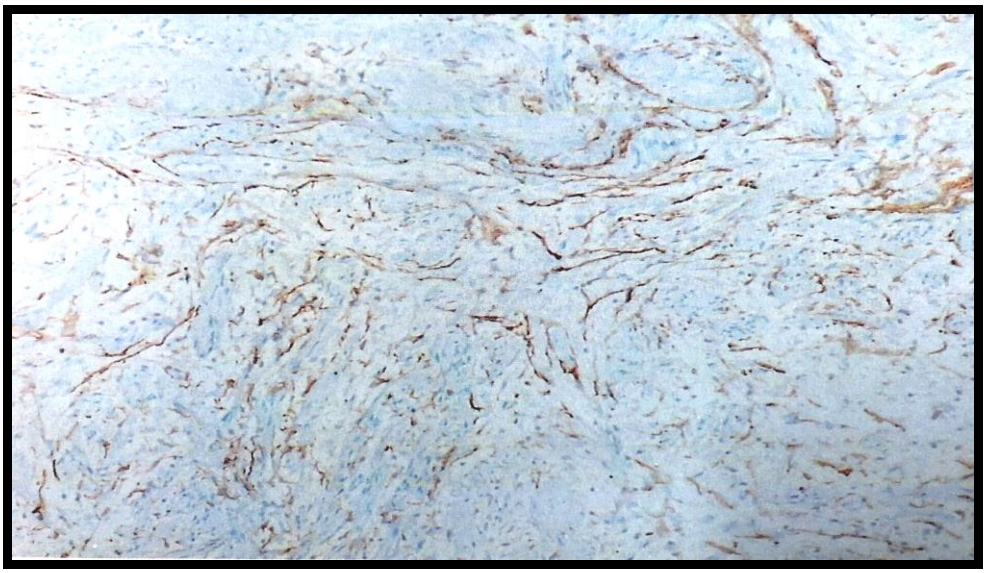

Figure 9: Tumor cells are negative for CD-10. (DAB, 10x).

DISCUSSION

NSM is a rare benign tumor of peripheral nerve sheath origin. It was first described by Harkin and Reed. Gallager and Helwig described a series of tumors under the designation of nerve sheath tumors.[5,6] It usually involves fingers and knees.[1] There have been misconceptions regarding the line of origin and its relationship with Neurothekeoma. According to recent studies, it is postulated that both the lesions are distinct entities based upon clinical, histopathological, immunohistochemical and genetic characteristics.[2,6] Sheth et al reported neurothekeoma have a peripheral nerve sheath origin.[6]

Neurothekeoma is more common than NSM and it usually occurs as a solitary lesion in the head and neck region.[7] According to reports, on gene expression profiling, all the subtypes of neurothekeoma resemble Fibrous histiocytomas originating from fibroblastic cells.[2,6]

Immunohistochemistry plays an important role in distinguishing neurothekeoma from NSM. NSM is diffusely positive for S100 and SOX-10 whereas neurothekeoma is negative for S100.

Histologically, Differential diagnosis includes other benign peripheral nerve sheath tumors and myxoid tumors.[5,7]

Myxoid NF occurs as an unencapsulated mass that lacks lobular architecture and show patchy S100 immunoreactivity.

Myxoid schwannoma lacks multinodularity and shows antoni A foci with verocay bodies. Cutaneous myxoma shows multinodular masses in the dermis and subcutis. On immunohistochemistry, it is negative for S100.[7,8]

Perineuromas show whorled fascicles in a myxoid or collagenized stroma and have strong positivity for EMA and is negative for S100.

Cellular and mixed neurothekeomas have lesser amount of myxoid matrix, more spindling and at times plump epithelioid cells, more anisonucleosis and greater mitotic activity than NSM. They are S100 negative.[4]

Complete excision with a free tumor resection margin is the treatment of choice for NSM. A close follow-up is recommended because of high local recurrence rates of upto 50%.[9]

CONCLUSION

We present a case report of NSM due to rarity and at an unusual location in the scalp. It is important to distinguish it from neurothekeoma as it has higher chances of local recurrences. Immunohistochemistry plays a pivotal role in arriving at a definitive confirmatory diagnosis.

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Conflicts of Interest

There are no conflicts of interest.

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