INTRODUCTION

Nerve sheath myxomas are rare benign tumors of nerve sheath origin. In 1969, Harkin and Reed[1] first described and coined the term “nerve sheath myxoma.” Previously, it was considered to be similar to neurothekeoma. However, recent studies have demonstrated that both the lesions have distinct origins. Nerve sheath myxoma is commonly seen in the dermis and subcutaneous tissue of extremities, especially of fingers. It is rarely seen over the head-and-trunk region.[2,3] In this article, we report a case of nerve sheath myxoma of the scalp.

CASE REPORT

A 55-year-old female patient presented to the surgical outpatient department with a superficial scalp swelling. On examination, the swelling was 2 cm in diameter and appeared to be soft, painless, and mobile [Figure 1]. The overlying skin was unremarkable. Based on the examination findings, a clinical diagnosis of lipoma was made and lesion was excised and sent to histopathology department.

Specimen received, on gross examination, showed a partly skin-covered nodular lesion measuring 1.5 cm × 1.0 cm × 0.5 cm. Cut surface of the lesion was gray-white to yellow.

Multiple sections were taken from the lesion and histopathological examination revealed a tumor comprising multiple variable-sized myxoid nodules surrounded by fibrous septa. The tumor cells were round to oval, spindle shaped with cytoplasmic projections against a myxoid background. Cytological atypia or mitosis was absent [Figure 2].

Based on these features, morphological differential diagnoses of nerve sheath myxoma and neurothekeoma were considered. The myxoid material in the tumor showed alcian blue positivity and periodic acid–Schiff stain negativity [Figures 3 and 4].

ABSTRACT

Nerve sheath myxoma, a superficial myxoid tumor, was first described in 1969 by Harkin and Reed. Tumor has cytological and histological resemblance with neurothekeoma, another cutaneous myxoid lesion. Nerve sheath myxoma affects individuals of all age groups and equal predilection for both genders with most favored sites being the fingers and knee. Here, we present a case report of nerve sheath myxoma of the scalp, diagnosed and confirmed with histopathology and immunohistochemistry.

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Immunohistochemistry panel comprising S-100, epithelial membrane antigen (EMA) and CD-34 was applied. The tumor cells showed S-100 positivity [Figure 5] and were negative for EMA [Figure 6] and CD-34 [Figure 7]. This helped in clinching the diagnosis of nerve sheath myxoma.

**DISCUSSION**

Nerve sheath myxoma is a rare benign, soft-tissue tumor. It was originally described by Harkin and Reed\(^1\) as a myxoid...
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benign tumor of neural origin arising from Schwann cells. It presents as a slow-growing asymptomatic nodular lesion in the dermis and subcutaneous tissues, the most common sites affected include the fingers and knees.[3]

Since the initial description of nerve sheath myxoma, there has been a considerable debate around its line of differentiation and its relationship with neurothekeoma. Recent studies have clearly differentiated these lesions as two separate entities based on histomorphological, immunohistochemical, and genetic parameters.[3,4]

On microscopic examination, nerve sheath myxoma reveals a well-defined multinodular tumor comprising myxoid nodules separated by fibrous septae. These nodules have loose clusters of benign spindle-stellate and epithelioid cells surrounded by myxoid stroma. The tumor cells are positive for S-100, glial fibrillary acidic protein (GFAP), vimentin, and collagen type IV.[9] Our case also showed spindle-stellate and epithelioid-shaped tumor cell in a myxoid background and showed positivity for S-100.

Sheth et al., in their gene expression profiling study, have suggested that nerve sheath myxoma has origin from peripheral nerve sheath cells, while neurothekeoma resembles cellular fibrous histiocytomas and they originate from fibroblastic cells.[3,4]

Immunohistochemically, neurothekeomas are positive for EMA and are negative for S-100 and GFAP and their most favored sites include head and neck, where it presents as a solitary mass.[6]

Other histological differential diagnoses of nerve sheath myxoma include myxoid tumors, including plexiform neurofibroma, myxoid neurofibroma, myxoid schwannoma, extraneural perineuroma, and cutaneous, juxta-articular, and intramuscular myxomas.[2] Myxoid neurofibroma is poorly circumscribed and an unencapsulated lesion, which lacks lobular architecture and shows patchy S-100 positivity.[7] Myxoid schwannoma shows foci of Antoni A with Verocay bodies. They do not show multinodularity but express S-100.[2] Extraneural perineuromas show a whorled fascicular pattern in a myxoid or collagenous stroma. They can be differentiated from nerve sheath myxoma on immunohistochemistry as perineuromas show strong positivity for EMA and are negative for S-100, GFAP, and CD34.[8] Cutaneous myxomas in the dermis and subcutis show multiple myxoid masses, with tumor cells, being spindle to stellate shaped. Cutaneous myxomas show CD34 positivity but are negative for S-100.[9] Intramuscular and juxta-articular myxomas might show similar histology but are clinically ruled out.[2] Plexiform neurofibroma, associated with neurofibromatosis, involves larger nerve trunks and is multinodular, myxoid lesions, which show S-100 immunoreactivity.[10] Table 1 summarizes the differences in immunohistochemical expression in myxoid tumors.[10] Table 2 compiles previous case reports of nerve sheath myxoma with clinical presentations and immunohistochemical profiles.

Table 1: Histopathological and immunohistochemical differential diagnosis of nerve sheath myxoma

| Tumor type            | Capsule | Myoid stroma | S-100 | EMA |
|-----------------------|---------|--------------|-------|-----|
| Nerve sheath myxoma   | +       | ++           | +     | +/- |
| Neurothekoma          | -       | +/-          | -     | +/- |
| Perineuroma           | +       | +/-          | +     | ++  |
| Schwannoma            | +       | -            | ++    | +/- |
| Neurofibroma          | -       | +/-          | +     | +/- |

Table 2: Details of the reported cases of nerve sheath myxoma

| Authors                  | Clinical features                             | Immunoprofile                                      | Immunoprofile |
|--------------------------|-----------------------------------------------|----------------------------------------------------|---------------|
| Bhat et al.[2]           | 32-year-old female, painless nodule over the neck | S-100-positive, EMA-negative                        |               |
| Malkoc et al.[2a]        | 32-year-old male, right paravertebral mass     | Immunoreactive for S-100, CD-10, and EMA. Negative for cytokeratin |               |
| Fathaddin and Fatanji[2b] | 23-year-old female, painless nodule over left thenar eminence | Tumor cells were positive for S-100 and vimentin, negative for GFAP, desmin, and CD-34 |               |
| Safadi et al.[3a]        | 32-year-old female with gingival swelling      | Positive for S-100, negative for EMA, GFAP, CD-68, SMA, and HMB-45 |               |
| Nakamura et al.[3b]      | 23-year-old female, swelling over the left lower eyelid | S-100 positive                                      |               |
| Bartake et al.[3c]       | 19-year-old female, growth at the anterior part of hard palate | S-100 strong positivity, NSE-focal variable positivity, and GFAP-negative |               |
| Spadari et al.[3d,e]     | 69-year-old male, nodular swelling over the tongue | Positive for S-100 and vimentin and negative for CD-34 and EMA |               |
| Present case             | 55-year-old female, painless nodular swelling over the scalp | Positive for S-100 and negative for CD-34 |               |

EMA – Epithelial membrane antigen; GFAP – Glial fibrillary acidic protein; NSE – Neuron specific enolase; HMB – Human melanin black; SMA – Smooth muscle actin

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We present a rare case report of nerve sheath myxoma, at an uncommon location of the scalp. It becomes important to distinguish nerve sheath myxoma from other myxoid tumors, especially neurothekeoma. Immunohistochemistry plays a pivotal role in diagnosis as histopathological features alone are not sufficient for the diagnosis. The differentiation from neurothekeoma is important as nerve sheath myxomas have a higher propensity to cause local recurrences.

**Informed consent**

Written informed consent was obtained from the patient for the publication of this case report.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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