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Case Report
Nerve Sheath Myxoma

Nerve Sheath Myxoma in an Uncharted Territory

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Dermal nerve sheath myxoma is a rare benign tumour of perineural Schwann cells of peripheral nerves. This is a case of an elderly female who presented with a swelling in the right nasal cavity. An excision biopsy of the mass was done and histopathology showed features of nerve sheath myxoma.

Keywords: NERVE SHEATH MYXOMA

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Introduction

Dermal nerve sheath myxoma is a rare benign tumour of perineural Schwann cells of peripheral nerves [1]. It commonly affects the limbs of younger adults with a peak incidence in the thirties [2] and has a high recurrence rate if not fully excised. Dermal nerve sheath myxoma is a benign peripheral nerve sheath tumour composed of small epithelioid, ring-like and spindled Schwann cells embedded within an abundant myxoid matrix. It typically arises in the skin or subcutis with a multinodular growth pattern. Tumors predominantly occur in the distal extremities with fingers being the most common site [3].

Occurrence of nerve sheath myxoma in the head and neck region is extremely rare. Histopathology and immunohistochemistry play an important role in diagnosis.

Case Report

A 70-year-old female presented with a slow-growing painless swelling in the right nasal cavity for one year (figure 1). Examination showed a non-tender ovoid pedunculated swelling measuring 2x1.5x0.5cm located in the right nasal cavity attached to the nasal vestibule inferiorly (figure 2). It was firm in consistency, freely mobile and not adherent to underlying tissues.

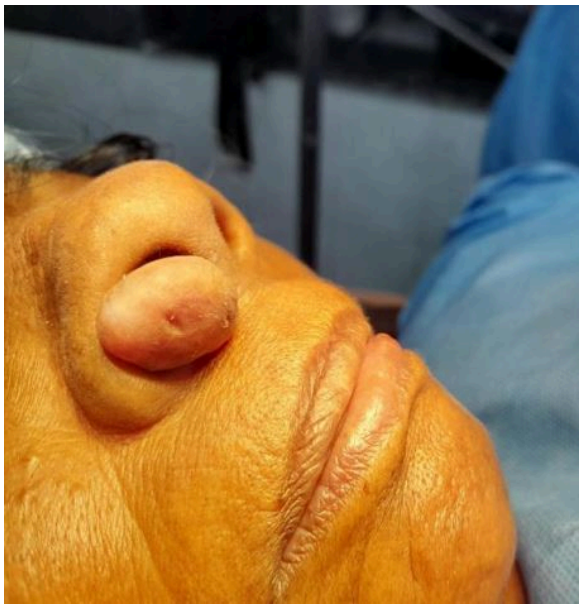

Figure 1: Mass protruding through the right nasal cavity.

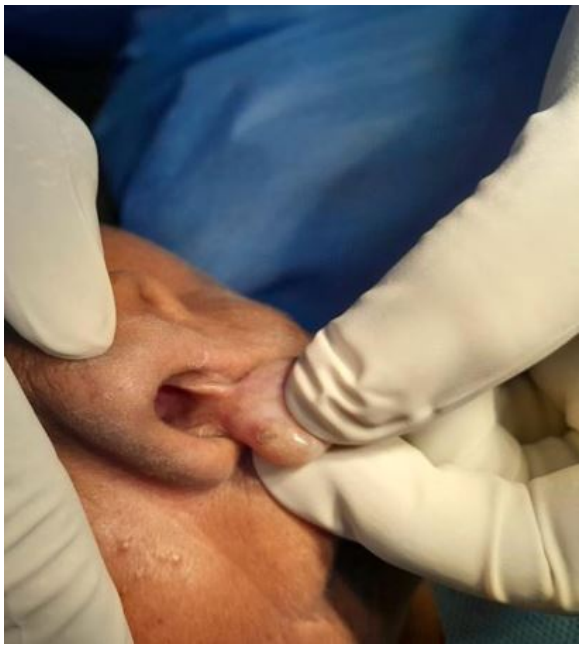

Figure 2: Nasal mass attached to the vestibule.

The lesion was excised and sent for histopathological examination (figure 3).

The Cut section showed gelatinous and yellowish areas with focal congestion. Histopathology showed skin with epidermis and an encapsulated neoplasm in the dermis with a multinodular pattern of cells separated by fibrous tissue. The cells were arranged in bundles and whorls within an abundant myxoid matrix.

Individual cells were spindle with some showing multi-polar cytoplasmic processes and some containing multiple vacuoles. No atypia or mitosis was seen. Excision margins were free. Immunohistochemistry was positive for S100, and vimentin and negative for cytokeratin. The patient was on regular follow-up for 6 months and there was no evidence of recurrence.

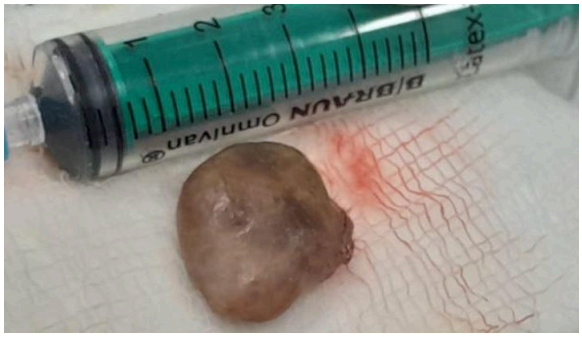

Figure 3: Nasal mass excision specimen.

Discussion

Nerve Sheath Myxoma (NSM) is a rare, benign Neuroectodermal tumour first described by Harkin and Reed in 19694. NSM predominantly occur in fingers and toes and its occurrence in the head and neck region is extremely rare. Dermal NSM typically present as small superficial growing masses, firm to rubbery in consistency. The lesions are usually asymptomatic, and rarely become painful [3] [4].

Neurothekeoma was considered a variant of NSM [5]. In 2011 Sheth et al., differentiated both based on genetic expression of cells using microarray analysis [6]. NSM was previously considered as the myxoid variant of Neurothekeoma [7], [8]. Standard imaging techniques have a limited role in the diagnosis of such a lesion. Histoimmunochemistry is the most accurate diagnostic tool. Wide local excision with a tumour-free surgical margin is the treatment of choice for NSM [9].

Conclusion

This is a rare case of nerve sheath myxoma in an uncommon location. The Nerve Sheath Myxoma and myxoid neurothekeoma were considered synonymous in the past but now using immunohistochemical markers, both can be differentiated. For Nerve Sheath Myxoma S100 is positive and is negative for neurothekeoma. Thus immunohistochemistry plays a pivotal role in arriving at the right diagnosis. This differentiation is important as NSM have a high propensity for local recurrence [10].

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