

*Case Report*

## Myxoid Neurothekeoma: A Case Report

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### ABSTRACT

Myxoid neurothekeoma is a rare benign cutaneous neoplasm of nerve sheath origin. It usually arise in childhood and early adult life. It can occur in skin, soft tissue or intraspinal location. Most commonly involves head, neck and shoulder. This rare tumor is misdiagnosed as other more common benign neuronal tumors. It should be distinguished from certain malignant tumors like fibrohistiocytic tumors or plexiform cell tumors, neurotrophic melanomas and clear cell sarcoma. This case report describes a rare case of neurothekeoma presenting as scalp swelling in a 29 year old female. Clinically, it was diagnosed as sebaceous cyst. Hence it is important to be aware of this entity to avoid misdiagnosis.

**Key words:** Neurothekeoma, benign neoplasm, nerve sheath origin.

### INTRODUCTION

Neurothekeoma is a rare benign cutaneous neoplasm of nerve sheath origin. It was initially described by Harkin and Reed in 1969 by name nerve sheath myxoma.<sup>[1]</sup> In 1980, Gallager and Helwig coined the term 'Neurothekeoma'.<sup>[2]</sup> It usually presents in childhood and early adult life. It can occur in skin, soft tissue or intraspinal location. Most commonly involves head, neck and shoulder.<sup>[3]</sup> This tumor shows overlapping features with other neural tissue tumors such as schwannoma, nerve sheath myxoma and neurofibroma, leading to difficulty in diagnosis.<sup>[4]</sup>

### CASE REPORT

A 29 year old female presented with painless scalp swelling since 3 years.

Physical examination showed a solitary, raised, slightly erythematous papule of firm consistency measuring 2x0.5x0.3 cm on scalp. Clinically it was diagnosed as sebaceous cyst. The mass was excised and sent for histopathological examination.

Gross examination revealed a skin covered nodular soft tissue piece measuring 2x0.5x0.3 cm the cut section was homogenous grey white.

On light microscopy, the H&E stained sections showed a multilobulated dermal lesion. The lobules were of variable sizes and separated by thin fibrous septa (Figure 1). Each lobule consisted of variable mixture of cells and myxoid stroma (Figure 2). The spindle and stellate cells showed abundant eosinophilic cytoplasm and oval nucleus with prominent nucleoli. A small

foci of slight atypia was also seen. On immunohistochemistry, the spindle cells were immunoreactive to S-100. They were, however, negative for GFAP (glial fibrillary acidic protein), desmin and CD34.

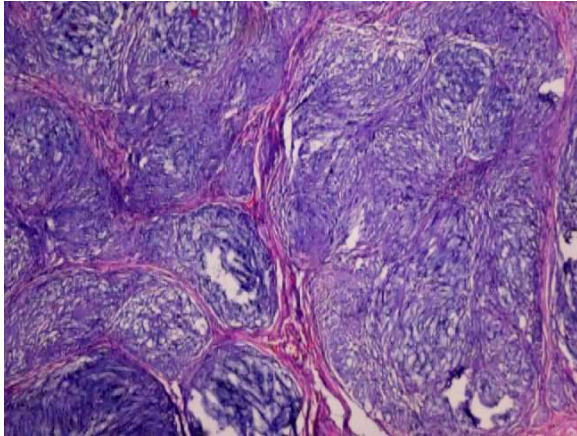


Figure 1: Lobules of variable size separated by fibrous septa. (H&E, 10X).

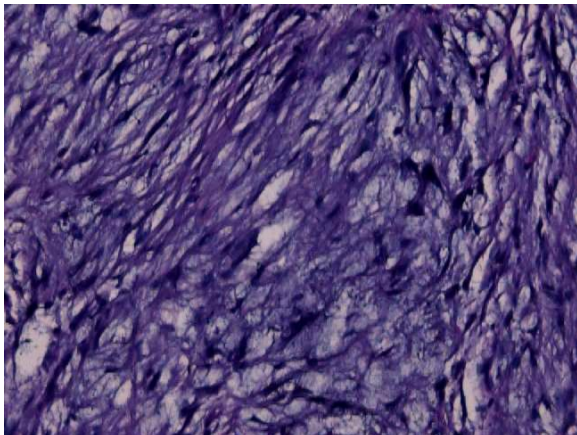


Figure 2: Lobule consisting of variable mixture of cells and myxoid stroma (H&E, 40X).

## DISCUSSION

Neurothekeoma is a benign tumor observed in young females mainly on face. It occurs as dermal cohesive mass without infiltration of epidermis. Based on histomorphological appearance and IHC findings, three variants have been described-myxoid, cellular and mixed types.<sup>[5]</sup> Myxoid type is characterized by greater degree of myxomatous change, less cellularity and well circumscribed spindle

cells in myxoid matrix and multinucleated giant cells. It is non encapsulated but well lobulated tumor. The cellular type is poorly circumscribed tumor consisting of nests and fascicles of epithelioid cells with vesicular nuclei containing scant myxoid stroma. Sclerotic collagen is more than myxoid type. These cells are S-100 negative and EMA (epithelial membrane antigen) positive thus distinguishing them from melanomas and showing its perineural origin.<sup>[5]</sup> The mixed or intermediate type show features of both cellular and myxoid variants.

Neurothekeomas must be distinguished from certain malignant tumors such as fibrohistiocytic tumors, plexiform cell tumors, neurotropic melanomas; neurofibroma, schwannoma and perineuroma. Schwannoma show nuclear palisading with proliferation of spindle shaped cells (Antoni type A). In myxoid neurofibroma, the myxoid matrix is seen only in limited area and is mixed with bundles of collagen. In neurothekeoma, myxoid is seen in all areas without collagen bundles.<sup>[6]</sup> Although in past, neurothekeoma has been reported as pacinian neurofibroma, cutaneous lobular myxoma, myxomatous perineuroma, but recently, markers like protein gene product (PGP) 9.5, NK1/C3 and S-100a6 have been proposed to be sensitive for identification of neurothekeoma.<sup>[7-9]</sup>

The treatment of choice is excision with clear margins and no recurrences or metastasis have been reported.<sup>[10]</sup>

## CONCLUSION

Although neurothekeoma is rare benign tumor, it should be considered in differential diagnosis of dermal lesions to distinguish it from malignant lesions so that aggressive therapy could be avoided.

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