Original Research Paper

Volume-8 | Issue-3 | March-2018 | PRINT ISSN No 2249-555X

Pathology

A CASE REPORT OF MYXOID NEUROTHEKEOMA OF THE SCALP - A RARE SOFT TISSUE TUMOUR

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ABSTRACT Neurothekeoma is a benign soft tissue tumour of neural origin. It is mostly seen in young females and commonly involves the head and neck region. We present a case of a 47 year old male, who presented with a nodule on his scalp. It was clinically diagnosed to be a case of sebaceous cyst however histologically proven to be a myxoid neurothekeoma. The histologic variants include myxoid, cellular and mixed tumors. Myxoid neurothekeoma is a cutaneous neoplasm of nerve sheath origin. This case report presents the rarity of the lesion that needs to be differentiated from lesions of similar histological features.

KEYWORDS : Neurothekeoma, myxoid, scalp.

INTRODUCTION:

Neurothekeoma is a benign soft tissue tumour of neural origin. It is mostly seen in young females and commonly involves the head and neck region. Here, we present a rare case of 47 year old male who presented with nodule in the scalp. Histology shows spindle cells set in a myxoid background. A diagnosis of myxoid neurothekeoma was made based on histological features. Neurothekeoma has to be differentiated from other spindle cell lesions. This case report is presented because of the rarity of the lesion and the need to be differentiated from lesions with similar histological features.

CASE HISTORY:

47 year old male presented with swelling in the scalp since 4 months. Examination revealed 2x2cm firm, nontender swelling in the frontal region of scalp.

Clinical diagnosis of sebaceous cyst was considered and the lesion was planned for excision under local anaesthesia.

Surgical excision of the lesion was done under local anesthesia.

On gross examination, there was a skin covered soft tissue, 2x1. 5x0.4cm. Sectioning showed a lesion in the dermis and subcutis, 1.3x1x0.4cm with a firm grey white to yellow cut surface. Histological examination revealed a lobulated tumour in the dermis composed of spindle shaped cells set in a myxoid matrix.

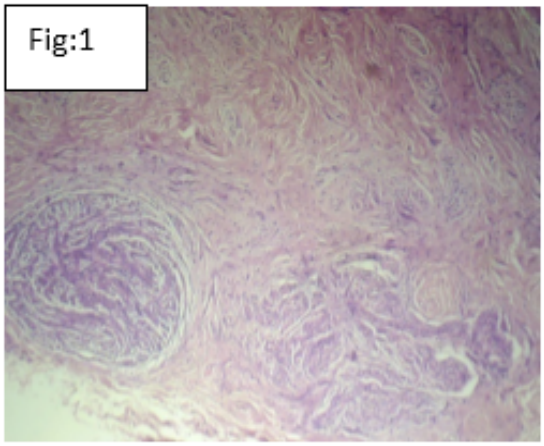

Fig 1: Histopathologic examination revealed a non-encapsulated tumor arranged in lobules separated by fibrous septae

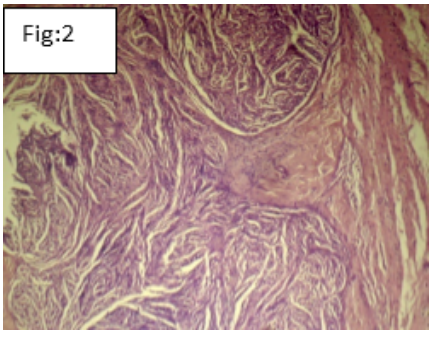

Fig 2: Non-encapsulated tumor arranged in lobular architecture separated by fibrous septae

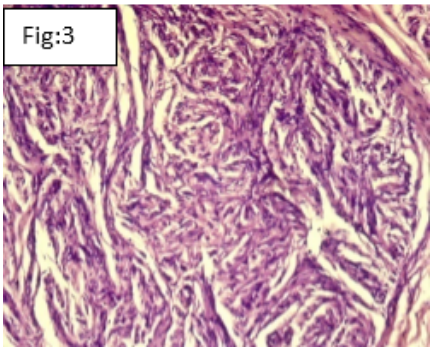

Fig: 3 Nests and bundles of spindle shaped cells set in a myxoid matrix

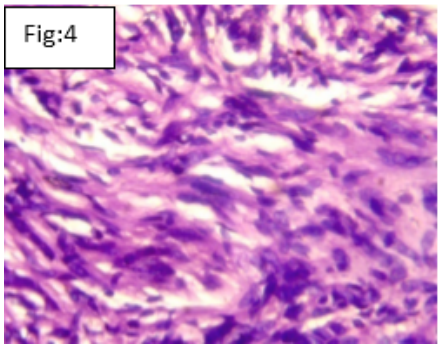

Fig: 4 Mild to moderate pleomorphism of epitheloid to spindle shaped cells with indistinct border

DISCUSSION:

The term "neurothekeoma" was first coined by Gallager and Helwig in 1980.[1] Neurothekeoma is a benign cutaneous dermal tumor.[2] It is composed of lobules of spindle shaped to stellate cells separated by fibrous septae. Neurothekeomas occurs in the first three decades of life with a female preponderance.[3] The common sites involved includes upper extremities and head and neck. Histologic variants include myxoid, cellular and mixed tumors.[4] Myxoid neurothekeomas are lobulated tumour located in the dermis composed of spindle shaped cells set in a myxoid matrix. Cellular neurothekeomas are poorly circumscribed tumours composed of epitheloid to spindled cells with abundant pale eosinophilic cytoplasm. Mixed neurotheokeomas shows histological features of both cellular and mixed types. Although the diagnosis of neurothekeoma can be made histologically, certain types like cellular neurothekeoma may require immunohistochemistry for confirmation. Neurothekeomas are immunoreactive for vimentin, NKI/C3, CD10, SMA, and CD68 and are negative for HMB-45, desmin, cytokeratin, S-100 and Melan A. [5] The differential diagnosis of myxoid neurotheokeomas includes neurilemmoma, neuromas , myxoid neurofibroma, mucosal melanoma and soft tissue myxoma. [4]

CONCLUSION:

Reporting rare cases like this will increase the awareness about this entity. These type of lesions should be included in the differential diagnosis of dermal lesion with myxoid stroma. Histopathology with immunostaining helps in diagnosing and differentiating various lesions with similar histology. Complete surgical excision is curative for these lesions.

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