

Benign Myxoid Dermal Lesion of Distinct Entity – A Rare Case Report



Medical Science

KEYWORDS : Benign neural lesion, nerve sheath origin, neurothekeoma, S100

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ABSTRACT

Neurothekeoma is a rare benign nerve sheath neoplasm also known as nerve sheath myxoma. It usually arises in childhood and early adult life. Neurothekeoma is commonly found in head, neck, shoulder and distal extremities. It has predominance for males. We report a case of neurothekeoma in a 52 Years old female with a single swelling in the scalp for the past 6 months which was clinically diagnosed as trichelemmal cyst. It is important to be aware of this entity to avoid misdiagnosis and delineate other myxoid lesions.

Introduction

Neurothekeoma is a rare benign cutaneous neoplasm of peripheral nerve sheath derivation. It was first described by Gallagher and Helwig.^[1] Before Neurothekeoma was coined, Neurothekeoma like cases had been reported as nerve sheath myxoma, dermal nerve sheath myxoma.^{[2][3]} Neurothekeoma usually arise during childhood and early adult life and have a predominance for men. It commonly involves head, neck, shoulder and distal extremities.^[4] They are situated in the dermis and subcutis but can occur in soft tissue and intraspinal locations also^[5]. Neurothekeoma shows overlapping features with other neural lesion such as schwannoma, myxoid neurofibroma, perineurioma, myxoid malignant fibrous histiocytoma leading to difficulty in diagnosing this entity.^[6]

Case Report

A 52 year old female presented with complaints of swelling in the scalp for a duration of 6 months which was gradually increasing in size. She had no other relevant complaints like pain, change of colour and ulceration. The physical examination showed a single smooth and dome shaped swelling measuring less than one cm. It was soft to firm in consistency. Clinically it was diagnosed as trichelemmal cyst. Excision of the swelling was done and sent for histopathological examination. Gross examination showed a skin covered soft tissue measuring around 0.8cm and the cut section was grey white. The Haematoxylin and Eosin stained sections was viewed under light microscopy which showed skin with underlying dermis composed of well defined multilobulated lesion of variable sizes separated by thin fibrous septa. (Figure 1). The lobules comprised of spindle and stellate shaped cells in a myxoid background. (Figure 2) Occasional multinucleated giant cells were seen. The diagnosis of Neurothekeoma was labelled based on distinct histology. However immunohistochemistry staining showed S100 positivity in the stellate cells (Figure 4) which helped to exclude cellular neurothekeoma and myxoid malignant fibrohistiocytic lesions.

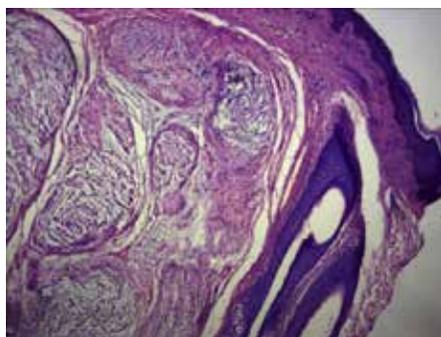


Figure 1: Skin with multilobulated dermal nodule of variable sizes separated by thin fibrous septa. (H&E, 4X).

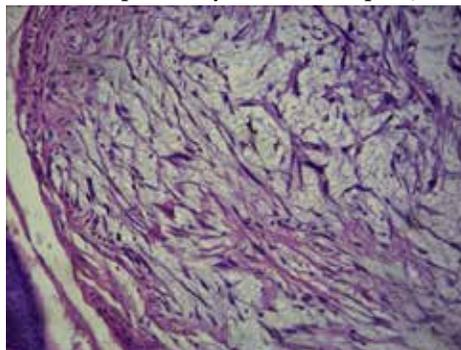


Figure 2: Lobules composed of oval-to-plump spindle and stellate cells, in a myxoid stroma (H and E, 10X)

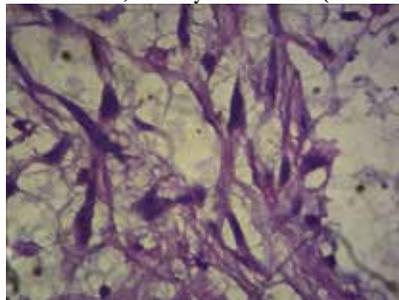


Figure 3: Spindle cells with nuclear atypia features not associated with aggressive behaviour. H&E 40X

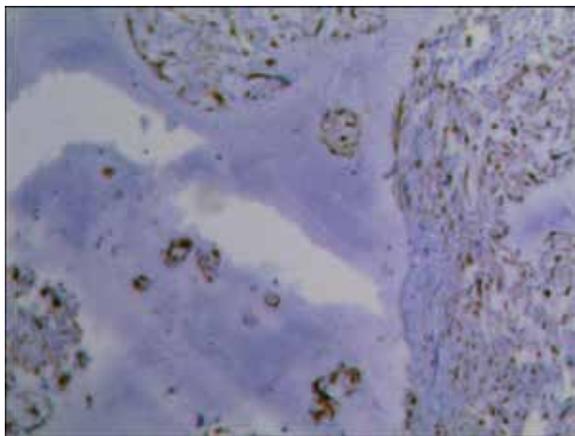


Figure 4: The tumor cells show positivity for S-100 immunohistochemical stain

Discussion

Myxoid neurothekeoma is a rare benign neoplasm of peripheral nerve sheath derivative. Clinically, neurothekeomas are slow growing and it typically presents as a small superficial asymptomatic, painless, firm, dome shaped nodule. This neoplasm usually arises during childhood and early adult life of young males with a predilection for head, neck and distal extremities. Mucous membranes are rarely involved. It is commonly seen in the dermis without involvement of the epidermis. Three histological variants have been described in Cutaneous neurothekeomas, they are Myxoid (classical or hypocellular), cellular and mixed types.^[7, 8] These tumors are benign and metastases have not been reported.^[9, 7] Thus, radiation and chemotherapy are not recommended as treatment for these neoplasms. The myxoid subtype of neurothekeoma is characterized by a lobulated, well-circumscribed proliferation of spindled and stellate cells in varying proportions within a myxoid stroma. Occasionally multi-

nucleated giant cells can be seen. The lesions characteristically stain positive for S-100 and PGP9.5^[10] is a broad neural marker but are negative for epithelial membrane antigen and markers of histiocytic differentiation. In contrast, the cellular types of neurothekeoma are not encapsulated and have an ill-defined fascicular growth pattern consisting of plump to epithelioid spindle cells, which may show nuclear atypia but lack stromal mucin and S-100 immunoreactivity.^[11] The third, mixed subtype shows features intermediate between classic and cellular types. The most helpful clues to the recognition of neurothekeomas are features like circumscription, superficial dermal location and distinctive septate architecture. Our case is a typical example of a myxoid type of neurothekeoma. The differential diagnosis of this rare tumor includes notably focal mucinosis, myxoid malignant fibrous histiocytoma, and myxoid neurofibroma^[1,13] clinical features, distinct histopathology and immunochemistry helps in diagnosing different variants of neurothekeoma and also differentiating other myxoid dermal lesions which has overlapping features leading to difficulty in diagnosis^[12]. Complete surgical excision with clear margins is the mainstay of treatment.^[9] But recurrence is common if excision is incomplete.^[14] Hence Neurothekeoma is regarded as a benign neural tumor and its recognition as an entity is important to avoid misdiagnosis of other aggressive lesions.^[1]

Conclusion

Neurothekeoma is a rare benign tumor which is simple enough to diagnose but should be considered in the differential diagnosis of other myxoid dermal lesions so that aggressive therapy could be avoided.

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