Myxoid Neurothekeoma – Case Report of An Unusual Nerve Sheath Tumour

G Mehdi, H Ansari, A Khan

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Abstract

Neurothekeoma and/or nerve sheath myxomas have been the subject of much discussion and research with varying opinions regarding their histogenetic origins. A careful evaluation of light microscopic features is required in these cases in order to exclude a malignant growth. In the following pages, we discuss the case of a patient with a nerve sheath myxoma clinically diagnosed as a ganglion.

INTRODUCTION

The terms ‘nerve sheath myxoma’ and ‘neurothekeoma’ both refer to benign tumours of presumed neural origin. Though the terminology is interchangeable, these tumours have a varied histomorphology and may possibly be of unrelated histogenesis. Those variants with a predominantly myxoid background are more aptly referred to as nerve sheath myxomas while the cellular forms are designated as ‘cellular neurothekeomas’. They are relatively rare lesions and the following report describes the case of a patient with the clinical diagnosis of a ganglion on the right ring finger.

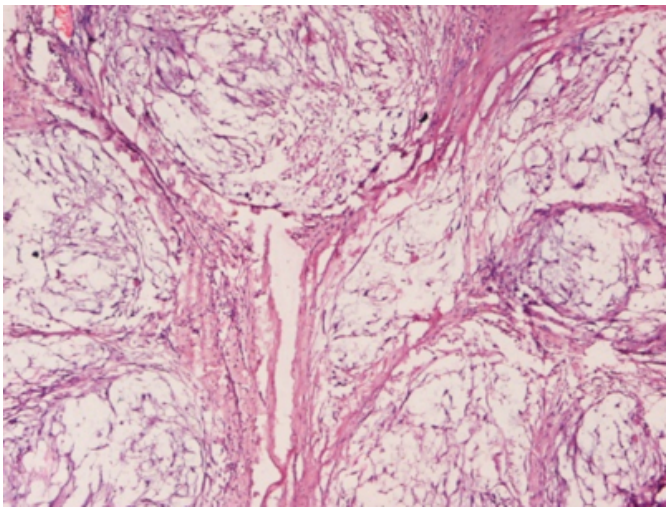
CASE REPORT

A 35 year old female patient presented with a growth on her right ring finger since three years. The clinical impression was that of a ganglion. After excision, the tissue was sent for histopathological evaluation. It appeared as a globular soft tissue piece, 2.5 cm in diameter, with a creamy white hue. The cut surface was mucoid, cream coloured and small cystic spaces were apparent on the surface.

On microscopic examination, the low power view revealed a multilobulated architecture. The lobules were separated by fibrous septations with a mononuclear cell infiltrate. Individual lobules were characterized by a prominent myxoid background and low cellularity (Fig.1).

Figure 1

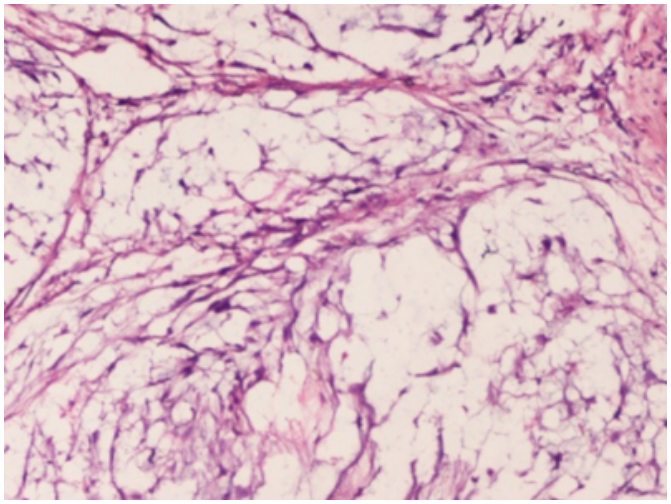
Figure 1: Lobulated architecture,myxoid background,low cellularity and intervening fibrous septae (H&E,×125).



The cells were present in small foci and were spindle / stellate in shape. Vacuoles were evident in several nuclei. In addition, anisonucleosis, focal nuclear hyperchromasia and occasional multinucleated cells were also present (Fig. 2)

Figure 2

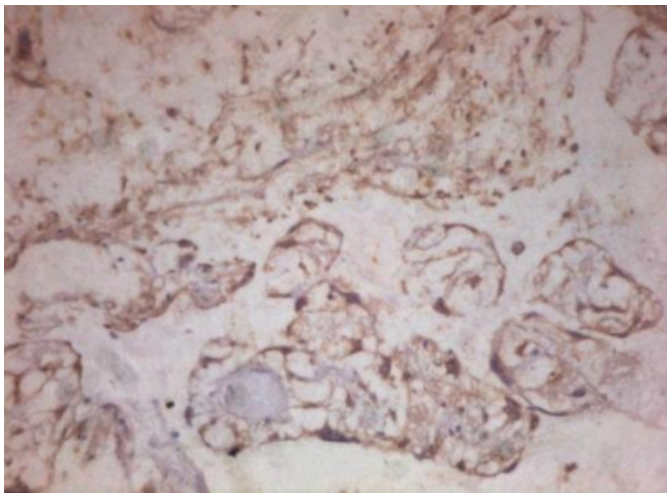
Figure 2: Scattered spindle cells with nuclear atypia and hyperchromasia (H&E,×500).



Mitotic figures were absent. The periphery of the lesion was demarcated by a fibrous capsule. The tumour showed S-100 positivity(Fig.3).

Figure 3

Figure 3: Myxoid neurothekeoma – S-100 positivity.



Keeping in view the described microscopic appearance, the diagnosis of a nerve sheath myxoma / neurothekeoma was considered appropriate.

DISCUSSION

The histogenesis of the dermal nerve sheath myxoma or neurothekeoma has long been debatable. They are generally sub -classified as tumours of nerve sheath or neural origin. It is now opined that the entity called neurothekeoma has three histologic variants-myxoid, mixed and cellular.[1] The 'cellular' neurothekeoma is distinguished by the paucity of myxoid areas and high cellularity.[1] Mitotic figures are common.[2]. These tumours also have a distinct immunohistochemical profile in contrast to the myxoid form. It is possible that the cellular neurothekeomas are of non – neural origin.[1]

In a retrospective studyon 22 tumours (neurothekeomas/ nerve sheath myxomas), it was observed that all myxoid variants showed reactivity for the S-100 protein, low affinity nerve growth factor receptor and variable affinity for glial fibrillary acidic protein (GFAP).[3] They were more common in males and had a peak incidence in the 4th decade. In contrast, both the cellular and mixed variants differed from the myxoid forms in morphology as well as in their negativity for nerve growth factor receptor. In addition, they were more frequent among females and peak incidence was in the 2nd decade. The authors of this study are of the view that the cellular and mixed variants do not show definite neurosustentacular differentiation. Other studies have also shown the cellular forms to be negative for S-100 protein[1] and positive for protein gene product 9.5 (PGP 9.5).[4] A recent hypothesis suggests its possible origin from fibroblastic cells which possess the capability of differentiating into myofibroblasts along with the acquisition of histiocytic cells.[5]

Clinically the lesion is commonly diagnosed as a ganglion. However, on histopathology, the differential diagnosis should include neurofibroma.[6] The typical neurofibrillary background of neurofibroma separates it from a nerve sheath myxoma, although myxoid change can cause confusion.[6] Differentiation from a fibrohistiocytic lesion can be sought by presence of circumscription in nerve sheath myxoma / neurothekeoma.[6]

An erroneous diagnosis of malignancy is also possible due to presence of nuclear atypia, hyperchromasia and mitotic figures. Occasionally, cellular neurothekeomas can mimic malignant melanoma.[2]. The presence of cells containing melanin has also been documented, which may pose a diagnostic dilemma.[7]

Important differentiating features of a cellular neurothekeoma are the presence of a well defined margin, lack of fibrosis and S-100 negativity.[2] Another clue to the diagnosis is the absence of a junctional / epidermal element in cellular neurothekeoma.[8]

In summary, the interesting morphological features of nerve sheath myxomas or neurothekeomas make them curious lesions which are apparently simple enough to diagnose but

require careful consideration of other lesions which can have an ominous prognosis.

CORRESPONDENCE TO

Dr. Hena A. Ansari 'Bait -Ul- Anwaar' Ahmad Gali, Medical Road AMU Aligarh – 202002 Uttar Pradesh, INDIA E-mail: hena.jnmc@gmail.com

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Author Information

Ghazala Mehdi, MD, DCH
Reader, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University

Hena A. Ansari, MD
Lecturer, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University

Arshad Hafeez Khan, MD, MCH
Reader, Department of Plastic Surgery, Jawaharlal Nehru Medical College, Aligarh Muslim University