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CASE REPORT

A Case Report on a Rare Entity: Nerve Sheet Myxoma

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Abstract

Nerve sheath myxoma is a rare benign tumor of peripheral nerve sheath origin arises from the Schwann cell. In 1969 Harkin and Reed first reported a case of Nerve sheath myxoma. The benign tumor clinically present as a nodular soft tissue swelling. Microscopic features are well circumscribed, varying sized nodules comprising of stellate cells in a myxoid background. The cells show diffuse positivity for S100. Nerve sheath myxoma to be differentiated from Neurothekoma, Myxoid Neurofibroma, Myxoid Schwannoma and Perineuroma.

Keywords: Peripheral nerve sheath tumour, Nerve sheet myxoma, Stellate cells, Neurothekoma, S100

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Introduction

Nerve sheath myxoma (NSM) is a benign tumour of peripheral nerve sheath origin, frequently found in extremities [1]. Incidence of nerve sheath myxoma was found more common in young adults [2]. The tumour commonly present as a non-tender, nodular lesion. The microscopic features are multiple myxoid nodule of varying size composed of stellate cells in a myxoid background. Previously nerve sheath myxoma and neurothekeoma were considered to be a related lesion but recent studied showed evidence of its discrete nature [3]. NSM show diffuse positivity for S100 and negative for EMA [4]. Differential diagnosis to be considered for NSM are neurothekeoma, myxoid neurofibroma, perineurioma and myxoid schwannoma [5].

Case History

A 2 years old girl presented with complaints of swelling in the right foot in 4th toe since one month. On Clinical Examination, the swelling was measuring 1.5 cm across, on the subcutaneous plane which was focally attached to the overlying skin. The swelling was soft, non-tender, non-pulsatile and slowly increased in size. X-ray examination showed well circumscribed, hypodense mass noted on the 4th toe of the right foot. Excisional biopsy was done. Grossly we received a well circumscribed, globular mass measuring 1.5x1x0.5cm. Cut surface homogenous, grey white with focal area showing mucoid material. Microscopic examination showed multiple well defined myxoid nodule of varying size which are separated by fibrous septae. The nodules were composed of stellate cells embedded in a myxoid stroma. Immunohistochemical marker S100 and EMA was performed, which showed diffuse positivity on S100 and Negative for EMA (Figures 1-3).

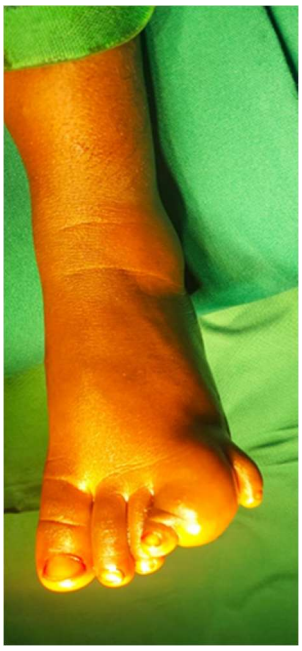

Figure 1. Show a soft tissue swelling measuring 1.5x1 cm on the right foot in 4th toe.

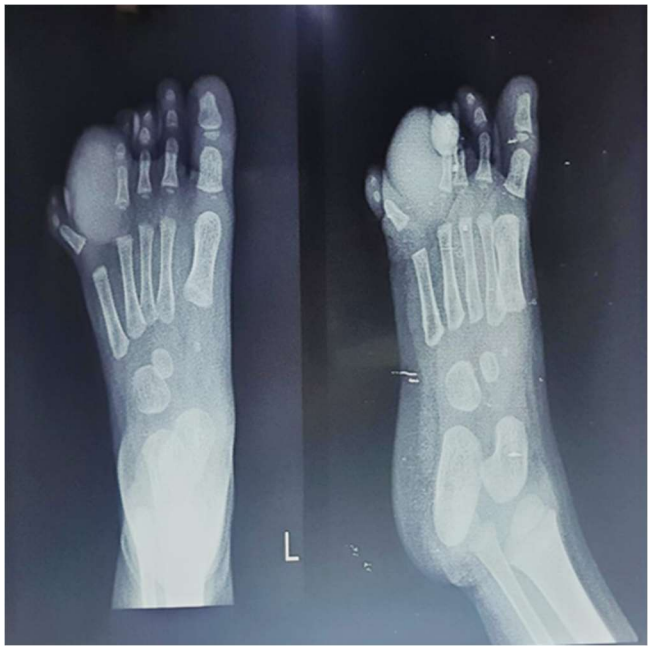

Figure 2. Show a well circumscribed hypodense mass.

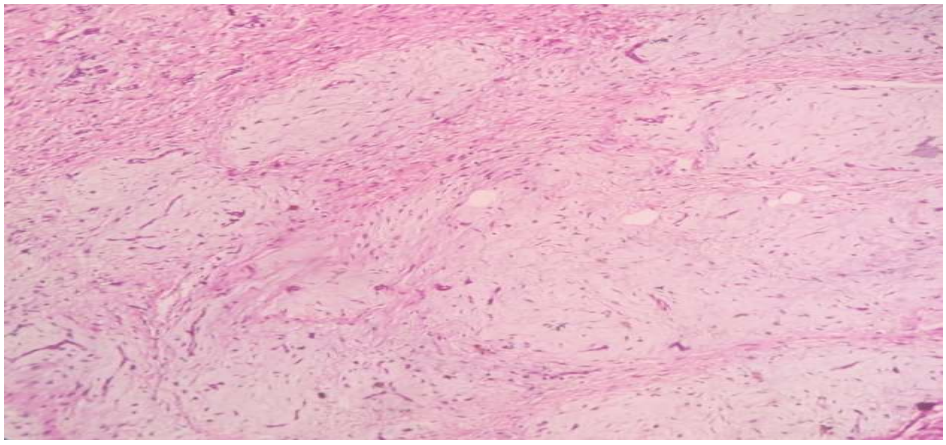

Figure 3. Shows multiple well defined myxoid nodule of varying size composed of stellate cells embedded in a myxoid stroma, which was separated by fibrous septae.

Discussion

Nerve sheath myxoma is benign tumor of peripheral nerve sheath origin arising from the Schwann cells. Nerve sheath myxoma was first reported by Harkin and Reed in the year 1969 [6]. Clinical presentation of nerve sheath myxoma is plainless nodular swelling with indolent growth pattern. Incidence of Nerve sheath myxoma was found in young adult with male to female ratio of 1:1 to 1:2 [7]. Commonest location of NSM is in the extremities with few cases have been reported in head and neck. Previously NSM and Neurothekeoma were considered to be a related tumor of peripheral nerve sheath origin but in recent

days, several studies showed evidence of difference between these two entities by means of clinical, histopathological features, immunohistochemical and genetic features [8]. Gross presentation of NSM is a well circumscribed, firm, multinodualr lesion. Cut surface grey white with myxoid areas. Microscopic features of NSM show a well-defined nodules of varying size which are separated by fibrous septae. The nodules are composed of stellate cells in myxoid background [9]. Immunohistochemical markers show positive for NSM are S100, GFAP and Negative for EMA. Differential diagnosis to be considered for NSM are Neurothekoma. Myxoid neurifibroma, Myxoid schwannoma and Perineuroma [10]. Nerve sheath myxoma need to be differentiate from neurothekoma by histomorphology and immunohistochemistry. Neurothekoma originate from fibroblast and resembles fibrous histiocytomas. It exhibit three different subtypes-cellular, myxoid and mixed. Neurothekomas are negative for S100 and positive for EM. Myxoid Neurofibroma is a unencapsulated tumor predominantly composed of spindle cells arranged in fascicles and whorls admixed with mast cells in a fibrillary and myxoid background. Neurites in Myxoid Neurofibroma show positivity for neurofilament protein. Myxoid schwannoma composed of spindle cells arranged in alternating hyper (Verocay A) and hypocellular (Verocay B) areas in a myxoid stroma. Perineuroma is a well demarcated, unencapsulated lesion composed of spindle cell (fibroblast-like) with variable degree of cellularity in a sclerotic stroma with foci of myxoid degenaeration.

Conclusion

In the present study we report a case of nerve sheath myxoma. Nerve sheath myxoma needs to be differentiated from other spindle cell tumors by clinical presentation, gross examination, histomorphology and immunohistochemtry. However Immunohistorychemistry play a vital role in diagnosing the spindle cell lesions and also guide us in arriving at the right diagnosis.

Statements and Declarations

Conflicts of interest

The authors declares that they do not have conflict of interest.

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