CASE REPORT

Benign nerve sheath myxoma: Atypical giant lesion in atypical location

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ABSTRACT

Benign nerve sheath myxoma (myxoid neurothekeoma) is a rare benign cutaneous neoplasm of nerve sheath origin. This tumor is commonly seen in the head and neck region and sometimes in the upper extremity. The presence of myxoid neurothekeoma on the lower limb is seldom and rarely reported. In this report we presented a new case of myxoid neurothekeoma in the lower limb of a 42-year-old male that showed a giant appearance.

INTRODUCTION

Benign nerve sheath myxoma [myxoid neurothekeoma (NT)] is considered one of the rare cutaneous neoplasm of peripheral nerve origin. It was first described in 1969 by Harkin and Reed as a micronodular, myxoid and loosely cellular benign tumour of neural origin.[1] Subsequently it was identified as neurothekeoma by Gallager and Helwig in 1980.[2]

Neurothekeoma commonly presents as an asymptomatic soft subcutameous nodule usualiy on face and upper extremities. It may present as a painful raised, skin colored, well-circumscribed lesion. It is more common in females of 21-36 years, ranging in sizes between 0.5-1cm. Clinically, a multitude of differential diagnoses exist for NT, including fibroma, dermatofibroma, leiomyoma, neurilemomma and neurofibroma.[3]

The cytological features of nerve sheath myxomas are less frequently documented in the past. However, the smears from nerve sheath myxomas show spindle -stellate cells and epithelioid cells with bland nuclei arranged in loose clusters, groups or whorls within a metachromatic myxoid background. Binucleated and multinucleated cells may be found.[4]

The histopathologic variants are: classic (myxoid) cellular and mixed types. Histologically classic (myxoid) hypocellular variant has a well-defined multinodular dermal lobules composed of myxoid nodules separated by fibrous septae, with nodules having loose clusters of benign spindled-stellate and epithelioid cells embedded in myxoid stroma. The cellular variant has an ill-defined fascicular growth pattern consisting of plump to epitheloid spindle cells, which may show nuclear atypia. The third, mixed subtype shows features intermediate between classic and cellular types.[5]

We report a case of large-sized myxoid neurothekeoma over the lower limb of an adult patient and review the literature for similar cases.

CASE REPORT

A 42-year-old man presented with slightly painful skin lesion on the left leg for 2 years. The lesion started as asymptomatic small nodule which slowly progressed and became slightly painful during the last few months. The patient

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gave history of exaggerated pain during touch or pressure on the lesion.

Physical examination showed a solitary, raised, slightly erythematous large nodule of firm consistency measuring about 1.4 cm in diameter [Fig. 1]. The lesion was located on the upper part of the anterior aspect of the left leg. It was slightly tender on palpation with no attachment to the underlying structures [Fig. 2]. X-ray studies showed a soft tissue density with no bony involvement.

The lesion was excised completely and gross examination showed multilobulated mass, measuring 1.4 X 1.5 X 1.2 cm in greatest dimension. The cut surface was tan yellow, with thin white septa dividing the mass into variably sized lobules.

Histopathological examination showed a multilobulated dermal lesion consisting of loosely arranged spindle and stellate cells within abundant stromal mucin [Fig. 3]. Some of the cells were multinucleated and had hyperchromatic, irregular nuclei. Mitotic figures were rare. Many cells had thin, delicate net-like projections imparting a sheaf-like configuration [Fig. 4]. Immunohistochemistry showed strong immunoreactivity of infiltrating cells with S-100 stain [Fig. 5]. There was no recurrence of the lesion after 6 months follow up.

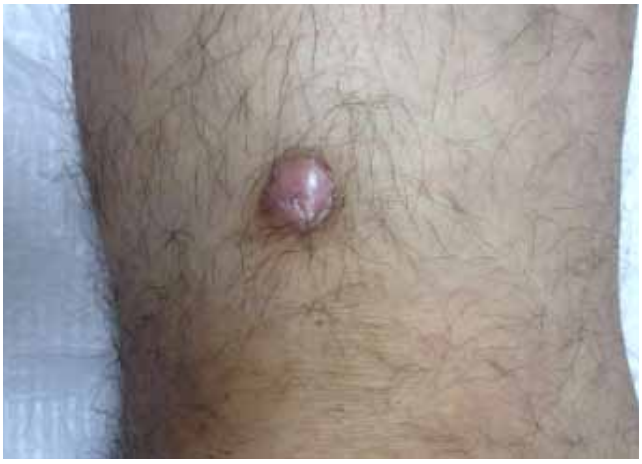

Fig. 1 A solitary large nodule on the anterior aspect of left leg.

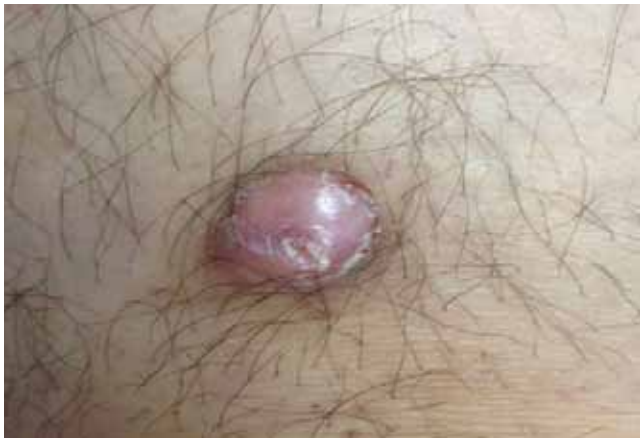

Fig 2 A well defined exophytic nodule with smooth glistening surface.

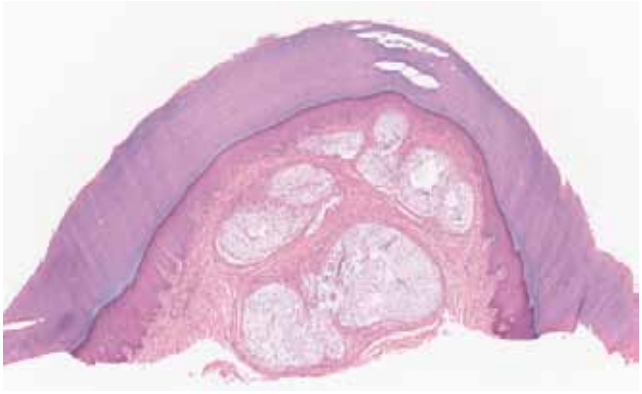

Fig. 3 A multilobulated dermal lesion consisting of loosely arranged spindle and stellate cells within abundant stromal mucin (H&E x40).

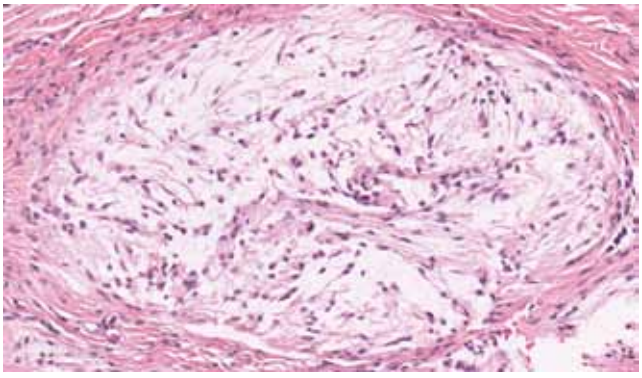

Fig. 4 Many cells had thin, delicate net-like projections imparting a sheaf-like configuration (H&E x400).

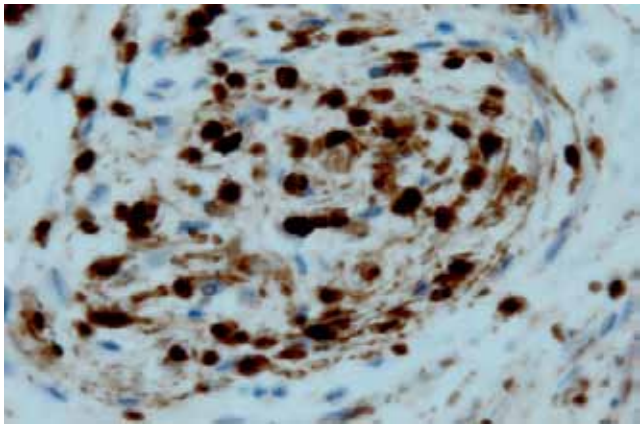

Fig. 5 Immunohistochemistry showed strong immunoreactivity of infiltrating cells with S-100 stain.

DISCUSSION

Neurothekeoma is a benign skin tumor that is usually related to a myxoid neoplasm of the soft tissue with presumed neural differentiation. The cytological differentials include nerve sheath myxoma, mixed neurothekeoma, myxoid neurofibroma, plexiform neurofibroma, myxoid schwannoma, extraneural perineuriomas, cutaneous myxoma, intramuscular and juxta-articular myxomas, cutaneous mucinosis, and hamartomas. All these lesions show myxoid material along with benign spindle to stellate cells on cytology. Histopathology and immunohistochemical examinations are extremely important in arriving at the right diagnosis. Because of rarity of other entities, spindle cell neoplasm probably myxoid neurofibroma was offered as the first cytological diagnosis.[6]

Amongst the few cases reported in the lower limb, foot,[7] hallux[8] and subungual location in the toe[9] have been described in literature. Another case reported in the ankle of a 62-year-old man showed extensive dystrophic calcification within the lobules of the tumor, which had prompted a misdiagnosis of foreign body granuloma.[10] Although NT is rare in the lower extremity, the clinician should consider this entity in the differential diagnoses, as it is imperative to distinguish it from malignant lesions so that aggressive therapy could be avoided.[11]

Three variants of nerve sheath myxoma were described based on histopathological evaluation. These subtypes included the myxoid type, the cellular type and the mixed type. The classical myxoid type is characterized by low cellularity and large amounts of myxoid matrix. The lesion is usually bordered and diffusely positive for S-100.[12] As in our case, the strong positivity of tumor cells for S-100, which is a well-established marker for Schwann cells and myelin sheath, favors the Schwann cell origin. The cellular types of nerve sheath myxoma typically are not well circumscribed, and the mixed type includes varied cellularity with focal myxoid regions. The differential diagnoses include schwannoma (also the malignant type), neurofibroma, neurilemmoma, leiomyoma, intramuscular myxoma and low-grade sarcoma.[13]

Reviewing the reported cases of nerve sheath myxoma, a consistent immunohistochemical profile of the three histopathologic variants has not been established. Schortinghuis[14] reported a myxoid case of nerve sheath myxoma which was negative for S100 while the myxoid nerve sheath myxoma cases reported by Nishioka[15] and Makino [16] were S100 and NSE positive. Penarrocha[17] and Nishioka et al.[15] reported two mixed-type cases, which were positive for both S100 and NSE. The diagnosis of the cellular variant was given for one of the three oral cases reported by Nishioka et al.[15] That cellular lesion was both S100 and NSE positive. The other two cases reported by Nishioka et al. were of classic and mixed variants and all

showed the same S100 and NSE immunoprofile.[15] Local excision is the treatment of choice for NSM. Recurrence is rarely reported after local excision. In summary, this case report describes the 6th reported case of lower limb nerve sheath myxoma and the first giant case presenting in the literature.

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