

*Case Report***Soft Tissue Perineurioma- A Distinct Peripheral Nerve Sheath Tumor:
An Under Recognized and Rare Entity**Ranjini Kudva^{1*}, Swati Sharma^{2*}, Neha Jain^{3*}, Seetharam Prasad^{4**}, Sandeep Kumar^{5***}¹Professor, ²Associate Professor, ³Post graduate, ⁴Associate Professor, ⁵Assistant Professor,
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*Received: 01/06/2015**Revised: 20/07/2015**Accepted: 02/07/2015***ABSTRACT**

Soft tissue perineurioma is a distinct peripheral nerve sheath tumor with characteristic immunohistochemical and ultrastructural features. It has a benign clinical course and complete surgical excision is curative. We describe this underrecognized and rare tumor in a 32 year old lady with its clinical, histologic and immunohistochemical features.

Key words: Perineurioma, peripheral nerve sheath tumor, immunohistochemistry

INTRODUCTION

Soft tissue perineuriomas (STP) are rare and under recognized benign peripheral nerve sheath tumors that exhibit perineurial cell differentiation. They were first described by Lazarus and Trombetta in the year 1978. ^[1] Usually subcutaneous in location but infrequently may affect deep soft tissues of the extremities or trunk as well. ^[2,3] STPs are characterized by slender spindle cells, arranged in short fascicles or whorls and focal storiform areas. Characteristic immunohistochemical (IHC) profile of perineurial cells include positivity for epithelial membrane antigen (EMA). ^[4] Acquaintance and awareness of this entity is important for correct diagnosis and also to avoid unnecessary aggressive local excision.

We report a case of soft tissue perineurioma and describe its distinct clinicopathologic features.

CASE REPORT

A 32 years old female came with the complaints of swelling in the left arm since 3 days. It was insidious in onset and gradually progressive. There was no associated pain, redness, fever or history of trauma. On local examination a 5x3 cms firm mass with regular borders was noted. Rest of the physical examination, hematologic and biochemical investigations were normal. Ultrasonography revealed oval hypoechoic lesion measuring 6x2.6 cms with rich internal vascularity located in the deep subcutaneous plane. Underlying bone was normal. Possibility of benign soft tissue tumor was suggested. Fine needle aspiration

cytology was inconclusive. Excision biopsy was done and sent for histopathologic examination. Gross examination revealed a single grey white glistening nodular tissue mass weighing 66 gms and measuring 6.5x5x1.5 cms. On cut section a well circumscribed lobulated mass with glistening white cut surface was noted (Fig 1). Microscopy showed a non encapsulated tumor composed of loosely spaced spindle cells with vesicular nucleus along with wavy streamer like cell processes arranged in storiform, whorled pattern and short fascicles pattern with sparse lymphocytic infiltrate and thin vascular channels (Fig 2). IHC was negative for S-100 and focal positive for EMA (Fig 3). Based on these findings a diagnosis of soft tissue extraneural perineurioma was given.



Fig 1- Lobulated mass with glistening white surface

DISCUSSION

Perineuriomas are rare peripheral nerve sheath tumors composed of well-differentiated perineurial cells which exhibit distinct ultrastructural and immunohistochemical profile.^[5] It accounts for approximately 1% of all soft tissue neoplasms. Perineuriomas are usually seen in middle-aged females.^[3] It may be intraneural or extraneural affecting the soft tissue. The extraneural type is further divided into a conventional form, and

variants like sclerosing and reticular types. Knowledge of these variants is important as they may form part of the differential diagnosis of other mesenchymal tumors.^[6,7] STPs are typically not associated with nerve and are usually benign. Nearly 70% are found in the superficial soft tissues. They have been reported to occur at rare sites like stomach, intestinal tract, kidney, lip, maxillary sinus, mandible, tongue and cardiac ventricle.^[3,8] STP usually presents as small, solitary, well circumscribed, non encapsulated, firm and painless nodule. Its size ranges from 1.5-7cm.^[3,4]

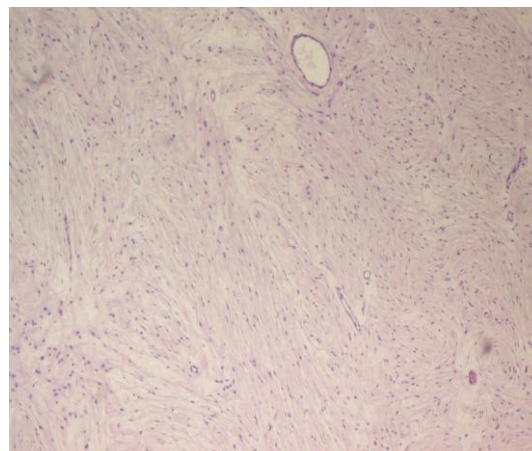


Fig 2- Spindle cells in storiform and whorled pattern H&E, X100

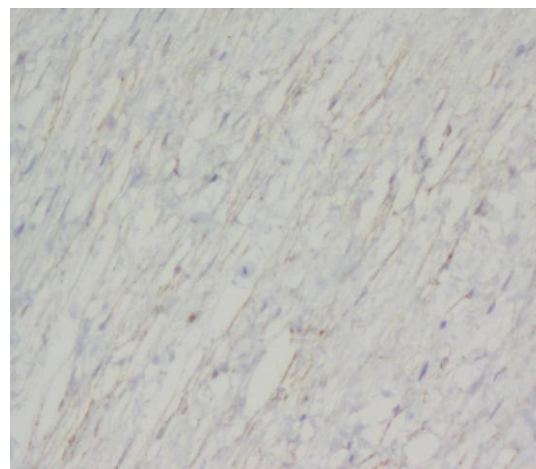


Fig 3- Focal positivity in tumor cells EMA, X2

The etiology of perineuriomas is not yet established. Possibility of being a reactive

lesion with proliferation of perineurial cells was ruled out as majority of the cases were not associated with trauma or associated inflammation. However, the perineurial proliferation has been shown to be clonal. Cytogenetically, STPs are associated with monosomy of chromosome 22 and loss of chromosome 13. Also loss of chromosome 10 and a small chromosome 22q deletion involving NF2 have been documented in literature. [3, 4]

On histology STPs are characterized by slender spindle cells, arranged in short fascicles, whorls, and focal storiform pattern. Nearly 50% of STPs are hypocellular whereas, 20% exhibit marked hypercellularity. The stroma varies from collagenous to a myxoid appearance. Around 40% of STPs show at least focal myxoid stroma, while 20% cases are exclusively myxoid (2). Nearly 20% of STPs may show atypical histologic features which include hyperchromatic nuclei, pleomorphic cells, and infiltrative margin. [3] On IHC, perineurial tumor cells are characteristically positive for EMA and negative for smooth muscle actin (SMA), S-100, and CD34. IHC profile helps in establishing the diagnosis of STPs. Our case showed typical histology and IHC features. No atypical findings were noted.

STPs have distinct morphologic, ultrastructural and immunohistochemical features. The histologic differential diagnosis include neurofibroma, schwannoma, smooth muscle tumors, solitary fibrous tumor, benign fibrous histiocytoma, extracranial meningioma, dermatofibrosarcoma protuberans and low grade fibromyxoid sarcoma. Lack of S-100 positivity excludes most peripheral nerve tumors with schwann cell component, but they are EMA negative as well which helps to differentiate them from STPs. Lack of staining for SMA rules out almost all the tumors with myofibroblastic or smooth

muscle origin. Solitary fibrous tumor cells stain negative for S-100 and EMA, and positive for CD34. In contrast, STPs do not show CD34 positivity. Benign fibrous histiocytoma stain negative for S-100, CD34 and EMA, and stain positive for CD68, hence EMA immunoreactivity for STPs will help to distinguish the two. The tumor cells of perineuriomas show more elongated, bipolar cytoplasmic processes compared to the meningothelial cells of extracranial meningiomas and tend to be arranged in fascicular or storiform pattern. Dermatofibrosarcoma protuberans have dermal origin with characteristic infiltrating pattern whereas, STPs are well circumscribed. Low-grade fibromyxoid sarcoma is composed of uniform bland spindle cells in alternating zones of collagenous or myxoid stroma with interspersed arcades of small blood vessels. Such alternating stroma is unusual in STPs. [2,3,4,7] Ultrastructural features of STPs include a discontinuous external lamina, junctional complexes, occasional pinocytotic vesicles and elongated spindled cell processes. [5]

The clinical course of STPs is almost always benign in nature. Local recurrence is extremely uncommon and reported in only 5% cases. [3,4] Complete surgical excision with a thin margin of normal tissue is curative. Atypical histologic features are believed to be a degenerative change and thus possess no clinical or prognostic significance. Malignant perineurioma also known as malignant peripheral nerve sheath tumors are characterized with infiltrative growth pattern, significant cytologic atypia, presence of necrosis and high mitotic activity. [8]

In conclusion, STPs are benign peripheral nerve sheath tumors composed of perineurial cells with characteristic immunohistochemical and ultrastructural features. Differential diagnoses include soft

tissue tumors composed of spindle cells. IHC and electron microscopy are helpful for confirming the diagnosis. STPs are frequently under recognized, hence awareness of this rare tumor is needed for its recognition and delineation of the clinicopathologic features.

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