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Extraneural Soft Tissue Perineurioma; A Rare Case Report

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Abstract:

Perineuriomas are rare benign peripheral nerve sheath tumors [PNSTs] exclusively composed of well-differentiated neoplastic perineurial cells. . Traditionally Perineurioma were thought to be neural only but the extraneural forms are also well known, infact more commonly documented. These are further subclassified into classical, sclerosing, and reticular/retiform subtypes. Here we describe a case of 30 year female with right thigh mass since four years and was diagnosed as a case of Soft tissue Perineurioma with classical morphology of bland spindle to oval cells with fibrillary cytoplasm arranged in vague storiform pattern and also forming whorls with a prominent perivascular arrangement (onion bulb formation) and characteristic IHC picture that was EMA positive and S 100 negative.

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Introduction:

Perineuriomas are extremely uncommon benign peripheral nerve sheath tumors exclusively composed of well-differentiated neoplastic perineurial cells[1]. About 200 cases have been reported worldwide[2]. It was first described by Lazarus and Trombetta in 1978 [3]. Later few case reports and a few large series followed[4]. Traditionally Perineurioma has been subdivided into intraneural (localized hypertrophic neuropathy) and extraneural/ soft tissue (further subclassified into classical, sclerosing, and reticular/retiform subtypes)[5,6,7]. Extraneural Perineuriomas are more common than Intraneural

Perineuriomas[8]. Here we describe a case of young female who presented with right thigh mass since four years and was diagnosed as a case of soft tissue Perineurioma.

Case Report:

A 30 year female presented to the Surgery OPD with a painless, right anterior thigh swelling that was present since four years with gradual increase in size. There were no other associated complaints. Her general condition was good and on local examination there was a diffuse firm swelling in the mid thigh measuring approximately 1x1 cm. After routine haematological investigations that were completely normal, the patient underwent excisional biopsy of the swelling and the tissue was sent to our department for histopathological examination.

Grossly multiple grey white pieces of tissue were received measuring 0.3x0.3 cm to 0.5 x0.5 cm and were processed completely .

Microscopically an encapsulated tumour was seen composed of bland spindle to oval cells with fibrillary cytoplasm arranged in vague storiform pattern and also forming whorls with a significant perivascular arrangement (onion bulb formation). At many places collagenisation was also noted. No significant mitosis, necrosis or atypia was seen. Based on light microscopic findings, a probable diagnosis of Perineurioma was made [Figures1-4].

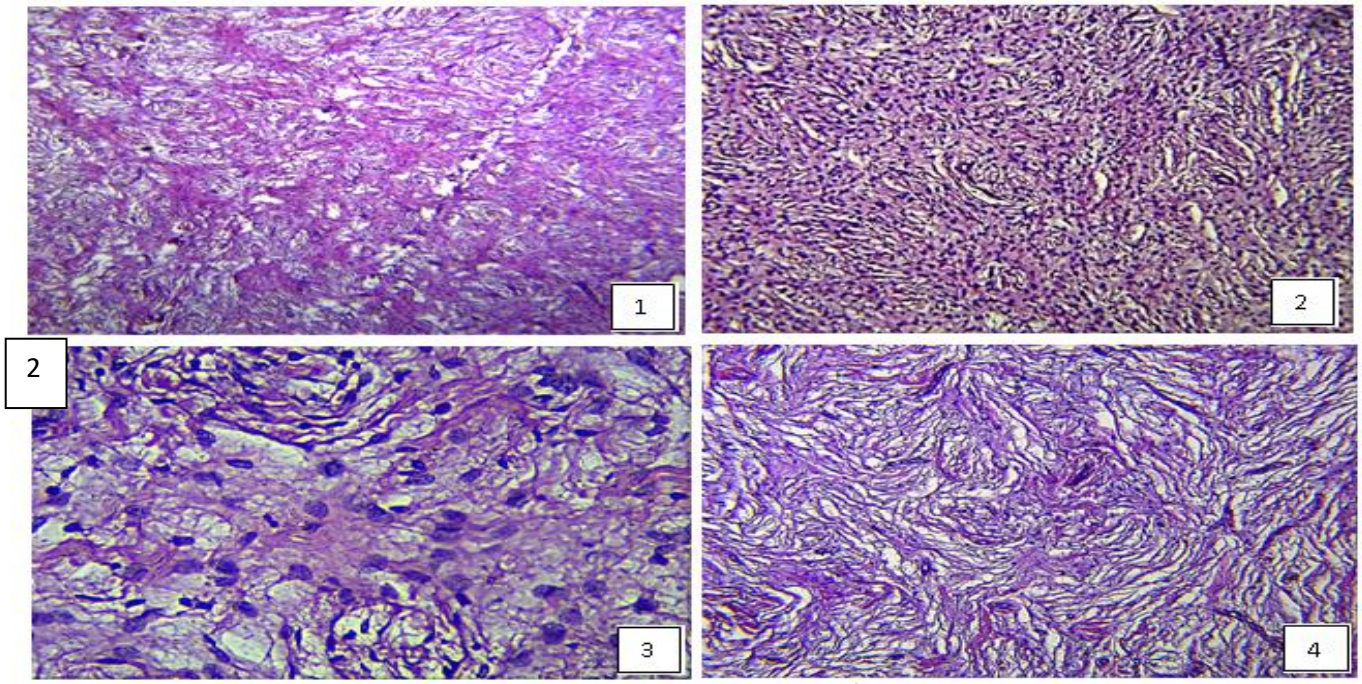

Figure 1-4 Perineurioma: (1) H and E 4x view showing storiform and whorling pattern. (2) H and E 10x view of the same. (3) H and E 40x view showing plump spindle cells. (4) H and E Characteristic perivascular whorling.

On immunohistochemistry, the tumor cells showed diffuse positivity for epithelial membrane antigen (EMA) but were negative for S100 thus confirming our diagnosis of Perineurioma [Figures 5, 6].

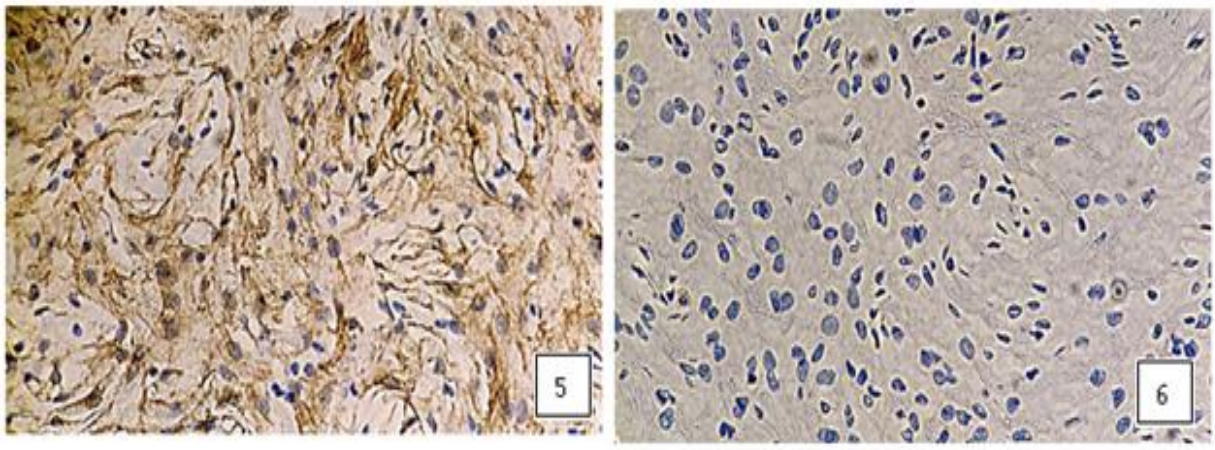

Figure 5-6 Perineurioma: (5) IHC EMA positive. (6) IHC S 100 negative.

Discussion:

Soft tissue Perineurioma is a very rare distinctive tumor entity with perineurial cell differentiation as evident from regular expression of perineurial cell markers and ultrastructural features of perineurial cells [4]. Pinkus and Kurtin in 1985 , first described EMA expression by immunohistochemistry in perineurial cells[9]. The ultrastructural features of perineurial cells are long thin cytoplasmic processes with numerous pinocytotic vesicles, abundant collagenous stroma, in-continuous basal lamina, and rudimentary intercellular junctions.[3]

Soft tissue[extraneual] Perineurioma arises typically in the soft tissue of adults and is more common in the superficial soft-tissue (70%) than in deep tissues. Though they show a predilection for the extremities and trunk [2], head and neck area, stomach, retroperitoneum, brain, kidney, maxillary sinus, mandible, and intestines are the other regions where these have been reported [10]. They are generally seen in young to middle-aged adults, more commonly in females, however the sclerosing variety is predominantly seen in males. [2]

Chromosomal aberrations affecting the neurofibromatosis type 2 locus (NF-2) at chromosome 22 have been reported in many cases of soft tissue Perineurioma [11,5] and somatic NF2 mutations are well known to occur in sporadic Perineuriomas [11,12], yet Perineuriomas are not perceived to be associated with NF-1 or NF-2, with the exception of a single case which was reported by Ausmus et al. in a 20-year-old patient with NF-1 [11,12].

It is a benign tumor with a low recurrence rate (5%) The usual presentation is as a painless, solitary, firm mass, that is generally not associated with an identifiable nerve [4].

Macroscopically, they are usually well circumscribed grey to white tumors, firm in consistency, varying in size from quite tiny to as large as 20 cm. Histologically, the tumors are unencapsulated, with microscopically infiltrative margins reported in 15% [4] They are composed of elongated spindle-shaped neoplastic cells , wavy-shaped nuclei with tapering ends and elongated, thin eosinophilic cytoplasmic processes forming whorls, lamellar or storiform arrangement with indistinct cell boundaries. Myxoid, collagenous, or hyalinized stroma may be seen. Mitotic figures and necrosis are usually absent.

Immunohistochemistry is required for accurate diagnosis of Perineurioma that stains with the perineurial cell markers- EMA, Claudin-1, and human erythrocyte glucose transporter-1 (GLUT-1), as well as CD34 and SMA(focal) but are negative for S-100 protein [2].

The most important differential diagnosis of neural Perineuriomas includes other common nerve sheath tumors like Schwannomas , Neurofibromas and low grade Malignant Peripheral Nerve Sheath Tumors[MPNSTs]. However, most cases can be differentiated on morphology alone or with the use of immunohistochemistry. The soft-tissue Perineuriomas can be either encapsulated or unencapsulated composed entirely of perineural cells in a predominant storiform pattern with a negative S-100 and positive EMA; the Schwannomas are encapsulated, show Antoni A and Antoni B areas, are positive for S100, and negative for EMA; Neurofibromas are unencapsulated , consisting of variable admixture of differentiated schwann cells, perineural like cells, fibroblast, mast cells and residual interspersed myelinated and unmylinated axons embedded in conspicuous extracellular matrix and show extensive

S100 positivity; [13,14] MPNST shows palisading monomorphic cells, large vascular spaces, perivascular plump tumour cells and geographic necrosis. But one has to be aware that hybrid PNSTs with two components have also been reported albeit very rarely , including hybrid Schwannoma/Perineuriomas, hybrid Neurofibroma /Perineurioma and hybrid MPNST/Perineurioma[15,16]

Distinction of soft tissue Perineurioma from other mesenchymal tumors such as Benign fibrous histiocytoma in benign lesions due to the predominant storiform architecture in both and from Malignant fibrous histiocytoma with myxoid change and Low-grade fibromyxoid sarcoma[17] amongst malignant lesions as some atypical cellular features such as focal hypercellular areas, pleomorphic/ multinucleated cells, or focally infiltrative margins may also be noted in a few cases of Perineuriomas is very important. These features are considered to be a degenerative change and, therefore have no clinical or prognostic significance with no malignant transformations reported so far in any perineurioma[18].

To conclude, Perineuriomas are rare tumours with a well documented Soft tissue counterpart. So a pathologist should be aware of this entity when reporting spindle cell neoplasms of soft tissue as it can easily be picked up by its typical morphological and immunohistochemical profile.

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This is a novel work as Perineuriomas are very rare and Soft tissue Perineuriomas though well documented is still a rare diagnosis.

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