Palisaded Encapsulated Neuroma (PEN)

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Citation

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Abstract

Palisaded encapsulated neuroma (PEN), while not an uncommon benign neural tumor of the dermis, is nevertheless often misdiagnosed clinically and histologically. A case of PEN of the skin is presented with a brief review of the literature. Distinguishing features from other benign neural tumors of the skin, in particular schwannoma and neurofibroma, are emphasized.

CASE REPORT

A 37-year-old woman presented with a 5 mm, smooth papule of the chin. The lesion was present for an unknown duration.

Figure 1

Figure 1. Low magnification, H&E stain.

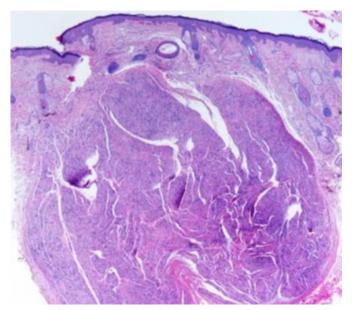


Figure 2 Figure 2. Higher magnification, H&E stain.

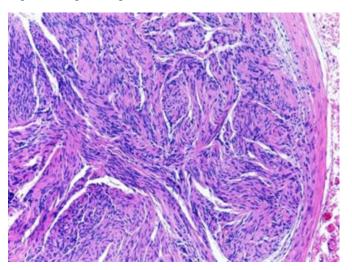


Figure 3

Figure 3. Tumor cells are strongly positive for S-100 protein.

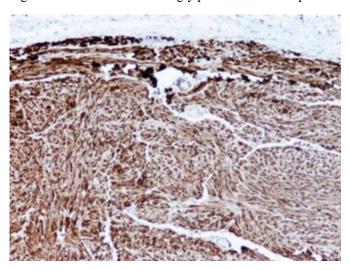
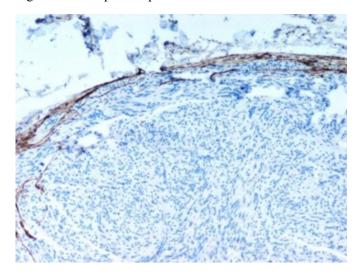


Figure 4 Figure 4. The capsule is positive for EMA.



Diagnosis: Palisaded encapsulated neuroma (PEN)

COMMENT

Palisaded encapsulated neuroma (PEN), otherwise known as solitary circumscribed neuroma, was first described by Reed et al in 1972 [1]. PEN, while accounting for approximately 25% of all nerve sheath tumors of the dermis [2], often goes unrecognized by the general surgical pathologist. Correct diagnosis is imperative, as misdiagnosis can result in unnecessary workup.

Clinically, PEN presents as a single, asymptomatic, flesh-colored papule, which usually appears on the face of an adult in middle age [3]. Often PEN appears close to a mucocutaneous junction, such as on the eyelid or near the lip [4, 5]. Appearance in other locations has also been reported

[1-4, 6]. There is a roughly equal sex predilection [6]. PEN is usually firm and gray-white [4]. These lesions may be mistaken clinically for basal cell carcinoma, melanocytic nevus, or fibroma [7], necessitating microscopic examination in establishing a diagnosis.

Microscopically (Figures 1 and 2), PEN is a well-circumscribed, partially encapsulated benign dermal tumor composed of interlacing fascicles of spindle cells. The fascicles are often separated by artifactual clefts created during specimen processing. The cells contain eosinophilic cytoplasm and tapered, wavy nuclei. Palisading of nuclei can be observed, but often is not prominent. Pleomorphism is absent and mitotic activity is minimal to none. The tumor is composed of S-100 positive Schwann cells and admixed neurofilament-positive axons. The capsule is composed of flattened, elongated epithelial membrane antigen (EMA)-positive perineural cells.

PEN can be confused histologically with other benign neural tumors, particularly schwannoma and neurofibroma [1, 2, 4, 5]. Neurofibroma lacks a capsule, contains mucopolysaccharide ground substance, and shows fewer axons within myelin sheaths [5]. In contrast to PEN, typical schwannoma is often located in the subcutaneous tissue, contains hypercellular Antoni A and hypocellular Antoni B areas with Verocay bodies, and lacks axons. Verocay bodypoor schwannomas exist, in which case the diagnosis becomes more challenging [8].

References

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