

Palisaded Encapsulated Neuroma

- Report of Two Cases -

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Palisaded encapsulated neuroma (PEN) is a distinctive benign cutaneous tumor that was first described by Reed et al. in 1972.

We have recently experienced two cases of PEN. Clinically, these lesions were asymptomatic, flesh-colored or reddish brown papules or nodules. Light microscopy revealed several lobulated, partially encapsulated dermal tumors composed of interlacing Schwann cell fascicles. In some areas, palisading of nuclei was observed, but this was not characteristic. On Bodian staining, moderate to large numbers of tiny axons were observed. Electron microscopy demonstrated many myelinated axons incompletely invested by Schwann cell cytoplasm.

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In 1972, Reed et al.1 described, for the first time, 44 cases of a distinctive cutaneous neural tumor that they designated "palisaded encapsulated neuroma" (PEN). Since that time, only a few convincing examples of this entity have been reported.2-5 Clinically, PEN is a slowly growing, asymptomatic, solitary, flesh-colored, firm papule or nodule usually located on the face of a middle-aged person.5 Histologically, it is characterized by a single or multiple encapsulated dermal mass of Schwann cells and numerous fine axons arranged in interlacing fascicles.1 The Schwann cells often show a characteristic nuclear palisading. Although PEN is not an uncommon tumor and has distinctive clinical and histological features, it is still very poorly known.

We herein present two cases of PEN that we have recently experienced.

REPORT OF CASES

Case 1. A 51-year-old woman visited our hospital with a slowly growing, solitary, asymptomatic, 5 mm-sized, flesh-colored, smooth-surfaced, dome-shaped nodule near the mucocutaneous junction of the upper lip for 1 year (Fig. 1). The patient's past and family history were not contributory. She denied any previous trauma history at the site of the lesion. On palpation, the nodule was soft and movable. Provisional clinical diagnoses included intradermal nevus, sebaceous cyst, and neurofibroma. The lesion was relatively easily shelled out by excision biopsy.

Histologic examination showed several well-circumscribed dermal masses composed of numerous spindle cells, which were partially surrounded by compressed bands of fibrous connective tissue (Fig. 2). Each tumor mass was composed of broad, elongated fascicles of Schwann cells. These fascicles were often separated from one another by clefts. Schwann cell fascicles were arranged in an interlacing pattern (Fig. 3). On Bodian staining, numerous tiny axons were demonstrated within the tumor mass (Fig. 4). Electron microscopy revealed many myelinated axons.
Fig. 1. A 5 mm-sized, flesh-colored, smooth-surfaced, dome-shaped nodule near the mucocutaneous junction of the upper lip in case 1.

Fig. 5. A myelinated axon (→) invested by Schwann cells with irregular nuclei (SN) and numerous lamellar processes in case 1 (Electron microscopy, ×20,000).

Fig. 6. A 3 mm-sized, reddish brown colored, firm papule (→) on the anterior thigh in case 2.

some of which were incompletely invested by Schwann cell cytoplasm (Fig. 5).

**Case 2.** The patient was a 9-month-old female infant having two reddish brown colored papules on the abdomen and thigh since birth. Cutaneous examination showed a 3 mm-sized, erythematous to brownish firm papule on the left anterior thigh (Fig. 6) and another 2 mm-sized lesion on the lower abdomen.

A biopsy specimen taken from the lesion of the thigh showed several lobulated, well-circumscribed dermal masses (Fig. 7). The dermal lobules were nearly spherical or ovoid in configuration. In the overlying epidermis, there was a mild degree of acanthosis, hyperkeratosis, and increase of melamin pigments in the basal cell layer. Numerous Schwann cells with deeply basophilic nuclei and ill-defined cytoplasmic outlines were arranged in interlacing fascicles and occasionally in a palisading pattern (Fig. 8). The Schwann cells were generally uniform, but occasionally showed some variation in the size and shape of their nuclei. The Bodian-stained tissue specimen revealed moderate numbers of fine axonal fragments oriented along fiber bundles within Schwann cell fascicles (Fig. 9).

**DISCUSSION**

Benign nerve sheath tumors are divided into three major categories: (1) neurofibromas, (2) neurilemmomas, and (3) neuromas. Neuromas can be divided into three types: (1) traumatic neuromas, (2) idiopathic solitary or multiple neuromas, and (3) multiple mucosal neuromas occurring in
Explanation of Figures

Fig. 2. A dermal mass composed of Schwann cell fascicles and encapsulated by a compressed band of fibrous connective tissue in case 1 (H & E stain, ×40).

Fig. 3. Magnified view of Fig. 2 showing Schwann cell fascicles arranged in an interfacing pattern (H & E stain, ×100).

Fig. 4. Numerous tiny axons (→) within the tumor matrix in case 1 (Bodian stain, ×100).

Fig. 7. Several lobulated, well-circumscribed dermal masses in case 2 (H & E stain, ×40).

Fig. 8. Magnified view of Fig. 7 showing that proliferating Schwann cells with deeply basophilic nuclei and ill-defined cytoplasmic outlines are arranged in interfacing fascicles and occasionally in a palisading pattern (H & E stain, ×400).

Fig. 9. Fine axonal fragments (→) oriented along fiber bundles in case 2 (Bodian stain, ×100).

multiple endocrine neoplasia, type IIb.

Idiopathic cutaneous neuromas were fairly rare and poorly documented until 1972. At that time, Reed et al. reported, for the first time, 44 cases of previously undescribed idiopathic cutaneous neuromas that they designated "palisaded encapsulated neuroma".

PEN is a slowly growing benign tumor that usually occurs on the skin of the face of middle-aged adults, but the lesion may develop elsewhere such as the glans penis or neck. The lesion may occur virtually anywhere on the face, but has a definite predilection for areas near mucocutaneous junctions. Clinically, the lesion usually appears as a solitary dome-shaped papule or nodule that is firm to palpation. The tumor in case 1 of this report was soft and movable, presumably because it was located in the relatively loose tissue of the upper lip. The tumors in case 2 were multiple-one lesion on the abdomen and the other on the thigh, and they resembled each other in size and shape. It is remarkable that they were present as multiple tumors and at birth, because all of the previously reported cases revealed a solitary lesion that had arisen in middle age to late adulthood. Unfortunately, we did not perform a biopsy of the other lesion on the abdomen.

Histologically, PEN is characterized by a single or multiple well-circumscribed dermal mass composed of compact Schwann cells and numerous tiny axons. Separate smaller nodules are often present in the dermis adjacent to the main portion of some of the tumors. These nodules are in continuity with the main portion of a tumor. The tumor mass is composed of narrow or broad, interlacing Schwann cell fascicles and is encapsulated, usually totally by a compressed band of fibrous connective tissue. The Schwann cells often show a characteristic nuclear palisading. Axonal stains such as Bodian stain show numerous axons in many of the fascicles. Electron microscopy demonstrates numerous axonal profiles, some of which are incompletely invested by Schwann cell processes. Histologic examination in our cases showed several lobulated, partially encapsulated dermal tumors composed of numerous Schwann cells and tiny axons. Our histopathologic findings are similar to those of Reed et al. and Dover et al. except in two respects. First, palisading of nuclei was not a prominent feature in our tumor, and it was observed only in some areas. Second, the tumor mass was only partially encapsulated by connective tissue bands. In fact, despite its original name, PEN does not really show typical nuclear palisading, a point also noted by others, and it is only rarely completely encapsulated.

Previously, the clinical diagnosis of PEN has been often mistaken for a basal cell epithelioma, intradermal nevus, or neurofibroma. The duration of the tumor, sparsity of telangiectasia, lack of a tendency to ulcerate, and well circumscribed location in the dermis help rule out the diagnosis of basal cell epithelioma. Intradermal nevi tend to be softer and to retain normal skin markings. PEN may be difficult to clinically differentiate from neurofibroma although PEN is firmer and has characteristic predilection sites.

The histologic differential diagnosis of PEN principally includes the other benign cutaneous neural tumors. PEN differs from neurofibroma in having the well-developed capsule and broader interlacing fascicles of numerous Schwann cells and abundant axons. Schwannoma is distinguished from PEN by its limited location in the subcutaneous tissue, the presence of Antoni type A and B tissue, and no demonstrable axons. Traumatic neuroma is also distinguished by its typical deeper location, a complete lack of circumscription, and the presence of either scarring or inflammatory cells.
Whether PEN is a neoplasm or the result of hyperplasia is a moot point, although Reed et al. prefer the latter explanation that the lesion is a primary result of hyperplasia of axons and Schwann cells.

As previously described, PEN is a not uncommon tumor that has distinctive clinical and histologic features. Therefore, when one encounters a firm papule or nodule near a mucocutaneous junction of the face in a middle-aged person, one should include PEN in the differential diagnoses.

REFERENCES