A Case of Solitary Circumscribed Neuroma

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Solitary circumscribed neuroma is a benign nerve sheath tumor of unknown etiology. Clinically, it appears as a long-standing, solitary, asymptomatic, skin-colored papule or papulonodule. It has been found predominantly on the face in both sexes, mostly in middle age. A 53-year-old female patient visited our department because of a nodule on the side of the distal interphalangeal joint of the left second finger. A biopsy specimen showed a well-circumscribed, partially encapsulated tumor composed of interlacing fascicles of spindle cells. Neither pleomorphism nor mitosis was found. Immunohistochemical stainings for vimentin, neuron specific enolase (NSE) and epithelial membrane antigen (EMA) were positive, but negative for S-100 protein and smooth muscle actin (SMA). We herein report an uncommon case of solitary circumscribed neuroma. (Ann Dermatol 12(1) 44-47, 2000).

Key Words: Solitary circumscribed neuroma

Although solitary circumscribed neuroma was first described in 1972 by Reed and colleagues as “palisaded encapsulated neuroma”, its etiology and pathogenesis have not been well clarified. This tumor is a benign cutaneous nerve sheath tumor. Clinically, the lesion appears as a slowly growing, solitary, firm, asymptomatic, skin-colored papule or nodule. The overlying skin is smooth and appears stretched. Occasionally, it is polypoid in appearance. It typically affects middle-aged patients and occurs with approximately equal frequency in both sexes. It appears almost exclusively on the face, but is also found in other sites, such as the shoulders, upper arms, trunk, hands and feet.

Herein we report one patient with the clinical, histopathological and immunohistochemical features of solitary circumscribed neuroma and review the literature concerning this subject.

CASE REPORT

A 53-year-old female presented at our depart-
Fig. 1. A solitary, 0.4 × 0.4 cm sized, well-circumscribed, round, skin-colored nodule with a smooth surface, which was found over the side of the distal interphalangeal joint of the left second finger.

Fig. 2. The tumor, located in the superficial dermis, was well-circumscribed and partially encapsulated by a band of parallel, flattened, elongated cells interspaced with collagen fibers (H & E stain, × 20).

Fig. 3. The tumor was composed of interlacing fascicles of spindle cells with poorly delineated eosinophilic cytoplasm and oval or wavy, deeply basophilic nuclei (H & E stain, × 160).

Fig. 4. Tumor cells stain strongly positive with antivimentin antibody (× 100).

Fig. 5. Neuron specific enolase (NSE) stained the nerve fascicles weak and focally (× 160).

Fig. 6. Staining for epithelial membrane antigen (EMA) was focally positive in the spindle cells of the tumor capsule (× 200).
DISCUSSION

Benign nerve sheath tumors are divided into three categories: schwannomas (neurilemmomas), neurofibromas and true neuromas. Neuromas can be divided into four types: extraneural (traumatic or acquired) neuromas, intraneural (isolated and spontaneous, solitary or multiple) neuromas, multiple mucosal neuromas occurring in multiple endocrine neoplasia type 2b, and abnormalities of sensory receptors. Idiopathic cutaneous neuromas are quite rare and asymptomatic. They may arise either in early childhood or in adulthood in the form of a solitary neuroma.

The solitary circumscribed neuroma is a long-standing, solitary, asymptomatic papule or papulonodule, found in both sexes, mostly in middle age and predominantly on the face or close to a mucocutaneous junction. No predisposing factor such as trauma is apparent. Local recurrence is an unusual phenomenon. The cause of solitary circumscribed neuroma is unknown. Reed et al. suggested that they might represent a form of multiple mucosal neuroma syndrome because of the histologic similarities and the proximity of the lesions to the mucosal surfaces. Dover et al. studied electronmicroscopic findings of the lesions and found the similarities to those seen in the regenerating axons. Therefore they suggested that the tumor might be traumatic in origin.

Despite its original name of palisaded encapsulated neuroma, solitary circumscribed neuroma does not really show typical nuclear palisading pattern noted by Dover et al., and it is rarely completely encapsulated. So, the term "solitary circumscribed neuroma" seems to be more appropriate.

Histopathologically, the tumor is located mainly in the dermis, but may on rare occasions extend into the superficial part of the subcutaneous fat. The surrounding dermis usually appears retracted from the tumor body. The tumor is usually separated from the normal appearing epidermis by a free zone of uninvolved papillary dermis. Rarely, it is in direct contact with the epidermis. The lesion is a well-circumscribed, partially or completely encapsulated tumor composed of interlacing fascicles of spindle cells with poorly delineated eosinophilic cytoplasm and oval or wavy, deeply basophilic nuclei. The fascicles are tortuous, of different thickness, and mostly separated from one another by artifactual clefts.

Immunohistochimical stains have been performed on many cases, and the results indicated that the immunoprofiles of the lesion were not specific and, in many respects, similar to those of the other cutaneous neural neoplasm such as neurofibroma, traumatic neuroma, etc.

Previous studies show that the lesion was positive for S-100 protein, NSE, and EMA in the tumor capsule. In contrast, our case revealed the lesion was negative for S-100 protein, but positive for EMA and NSE. We could not explain why staining for S-100 protein was negative in our case, but error in the procedure of staining, cutting unsuitable specimen, and other unknown causes could be possible. So, we should cut and stain the specimen again several times.

Solitary circumscribed neuroma should be differentiated from the schwannoma (neurilemmoma) or neurofibroma. Schwannomas classically have a biphasic appearance with Antoni A and B areas, frequently showing prominent nuclear palisading, and are completely encapsulated. Axons or nerve fibers are not found within schwannomas, other than occasionally in the capsule. By contrast, neurofibromas are unencapsulated, are usually comparably hypocellular with a loose rather myxoid matrix, and contain scattered small nerve fibers rather than innumerable tiny axons. Traumatic neuromas can be distinguished by their typically deeper location, a complete lack of circumscript, and their composition of innumerable, readily identified nerve fibers set in a collagenous stroma containing fibroblasts and perineural cells. The other rare consideration is angioleiomyoma, which arises very rarely on the face, contains prominent thick-walled vessels, and is composed of typical smooth-muscle cells with copious eosinophilic cytoplasm and blunt-ended nuclei.

We herein report an uncommon case of solitary circumscribed neuroma.

REFERENCES