A Case of Palisaded Encapsulated Neuroma of the Lower Lip

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Palisaded encapsulated neuroma (PEN) is a clinically-distinctive, benign, cutaneous nerve sheath tumor. It usually occurs as a solitary, asymptomatic, skin-colored papule, and commonly affects the face of middle-aged adults. We report a case of PEN which developed on the lower lip of a 44-year-old Korean man. It was a 5 mm-sized skin colored papule. Histopathologically, it consisted of uniform, broad, interlacing fascicles of spindle cells and was surrounded by a complete capsule in the dermis. On immunohistochemical staining, the tumor cells of the nodule were positive for S-100 protein and neural filaments, while the capsule of the nodule was positive for epithelial antigen.

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Key Words: Neural filament, Palisaded encapsulated neuroma, S-100 protein

INTRODUCTION

Palisaded encapsulated neuroma (PEN) was initially described and named in a series of 44 cases in 1972 by Reed et al. The lesion appears as a slowly-growing, asymptomatic papule, most often on the face. However, it is rarely found on other sites, such as the trunk, shoulder, upper arm, or mucocutaneous junctions. Only four cases have been reported in the literature so far in Korea. We present a case of PEN confined to the lower lip.

CASE REPORT

A 44-year-old Korean man presented with a 5-year history of an asymptomatic skin-colored papule on the lower lip. It had slowly increased in size. There was no history of trauma. He had no other significant medical history or notable family history. Physical examination revealed a slightly-raised, flesh-colored papule, which was freely movable and measured 0.5 × 0.5 cm (Fig. 1). The initial clinical diagnosis was a mucocele versus squamous cell carcinoma. An excisional biopsy was carried out and it was enucleated from its dermal bed.

A histological examination of the excisional biopsy revealed a well-circumscribed, round tumor in the dermis. It was composed of uniform, broad and interlacing fascicles of spindle cells. The fascicles were separated from one another by artifactual clefts. There was a focal tendency for the nuclei to align in a vaguely parallel fashion (Fig. 2A), but no evidence of classic palisading or Verocay body formation. The tumor cells had poorly-delineated, eosinophilic cytoplasm and their nuclei were wavy, rather pointed, and basophilic, but showed no pleomorphism (Fig. 2B). On immunohistochemical staining, the tumor cells were consistently intensely-positive for S-100 protein (Fig. 3A). The capsule was composed of flattened, elongated cells that showed positivity for epithelial membrane antigen (EMA) but the staining for EMA was negative within the body of the tumor itself (Fig. 3B). Immunoreactivity for neural filaments was detected focally between the spindle cell fascicles (Fig. 3C).
DISCUSSION

Histopathological findings of PEN revealed a well-circumscribed dermal nodule, composed of fascicles of spindle-shaped cells showing variable and focal nuclear palisading. Contrary to what the name implies, PEN does not really show typical nuclear palisading and it is only rarely completely encapsulated. The capsule was composed of flattened, elongated cells that showed positivity for EMA. The positivity of capsule cells for EMA may indicate that the capsule of these tumors originates from the perineurium, because antibodies to EMA are now known to decorate perineural cells. Our immunohistochemical findings were consistent with those of previous studies. The tumor cells were positive for S-100 protein and negative for EMA. In contrast, the cells of the tumor capsule were negative for S-100 protein and positive for EMA. The lack of EMA-positive cells within PEN differentiates it from traumatic neuroma, and the presence of axons labeled with neurofilament distinguishes it from Schwannoma.

PEN occurs almost exclusively on the butterfly area of the face, but may also involve other sites including the shoulder, upper arm, and the trunk. PEN is relatively rare on the mucosa. Reviewing the English literature, we found about 10 cases affecting the lips; 6 cases occurred on both lips, 3 cases on the upper lip, and one case on the lower lip. Four cases of PEN have been reported in Korean literature; two cases located on the cheek, one on the ala nasi, and one on the lower lip. We present an additional case occurring on the lower lip (Table 1). This condition appears to be rare on the lower lip.

The cause of PEN is a topic of considerable debate. Electron-microscopic findings are similar to those seen in peripheral nerve regeneration, suggesting that PEN may be traumatic in origin, and could represent regeneration following local minor
Table 1. Reported cases of PEN in Korean dermatologic literature

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age/Sex</th>
<th>Duration</th>
<th>Site</th>
<th>Skin lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hong et al.</td>
<td>70/F</td>
<td>2 years</td>
<td>Rt. cheek</td>
<td>Pea-sized, dome-shaped papule</td>
</tr>
<tr>
<td>Hong et al.</td>
<td>57/F</td>
<td>5 years</td>
<td>Rt. ala nasi</td>
<td>1cm-sized, polypoid nodule with telangiectasia</td>
</tr>
<tr>
<td>Park et al.</td>
<td>22/F</td>
<td>4 years</td>
<td>Rt. cheek</td>
<td>Pea-sized papule</td>
</tr>
<tr>
<td>Cho et al.</td>
<td>33/M</td>
<td>3 years</td>
<td>Lower lip</td>
<td>0.5 × 0.5 cm sized, dome-shaped papule</td>
</tr>
<tr>
<td>Present case</td>
<td>44/M</td>
<td>5 years</td>
<td>Lower lip</td>
<td>0.5 × 0.5 cm sized, slightly-elevated papule</td>
</tr>
</tbody>
</table>

injury to the skin. It has also been suggested that the classic form of PEN has a different histogenesis than traumatic neuroma, but chronic minor traumas could not be excluded as an etiologic factor of PEN on histologic grounds. Another study supports the fact that PEN is a reactive lesion rather than a true neoplasm. The involvement of the lower lip might not be as rare as reported in literature, considering that the lip is a site of various traumatic insult.

REFERENCES