Spiny Keratoderma on the Digit

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A few cases of a dermatosis characterized by “music box spine” keratotic papules limited to the palms and soles have been reported. There have been several names for this dermatosis. The most commonly incorporated term is porokeratosis, but Osman et al suggested the name “spiny keratoderma of palms and soles” because the clinical, histologic, and electron microscopic features of this lesion are distinct from porokeratosis. We report a case of spiny keratoderma on the digit showing a single lesion, as a variant of the classic spiny keratoderma of palms and soles. (Ann Dermatol 7(2):186-188, 1995)

Key Words: Porokeratosis, Spiny keratoderma

The dermatosis of fine, acuminate, quadangular, keratotic palmpoplantar papules resembling the spines of an old-fashioned music box cylinder has been designated by numerous terms, including “Punctate keratoderma,” 1 “Punctate porokeratotic keratoderma,” 2,3 “Porokeratosis punctata palmaris et plantaris,” 4,5 “Palmoplantar keratosis acuminate,” 6 “keratosis punctata palmaris et plantaris,” 7 and “spiny keratoderma of palms and soles.” 8 There has been much confusion as to whether this dermatosis is a form of porokeratosis or keratoderma. Osman et al concluded it as a form of keratoderma based on clinical and histologic evidence. Because malignant tumors can develop in porokeratosis, proper classification is required.

CASE REPORT

A 65-year-old man had an asymptomatic acuminate, keratotic papule on his left 3rd finger ofr 5 years(Fig. 1). The family history was noncontributory. He had tolerable gouty arthritis for 5 years, receiving no treatment for this disease. He had no history of hyperhidrosis or hypohidrosis. He was unaware of any exposure to arsenic. On physical examination, a single, discrete, firm, 1 mm, seedlike keratotic plug similar in appearance to a music box spine was seen on the lateral aspect of the digit. Microscopic examination of an excisional biopsy specimen(Fig. 2) demonstrated a central hyperkeratotic, parakeratotic column, so called cornoid lamella-like, with a reduced granular layer, and slight depression of the stratum malpighi beneath the parakeratotic column. No dyskeratosis or vacuolization was seen. The dermis and surrounding epidermis were normal. After excision of the lesion, no new lesions developed.

DISCUSSION

A few cases have been reported of lesions resembling the spines of an old-fashioned music box, limited to the volar aspects of the hands and feet, which were histologically composed of a column of parakeratotic material. Brown 1 first designated it as “puncatat keratoderma” and Herman 2 reported a case designated as “punctate porokeratotic keratoderma”, because he observed sweat ducts within and between the parakeratotic columns. But Lestringant and Berge 3 named it “Porokeratosis punctata palmaris et plantaris,” although it was distinct from true porokeratosis. Sakas and Gentry 4 classified these accumulate lesions as “Punctate porokeratosis” because the parakeratotic column was cornoid lamella-like. Cornoid lamellation is characterized by a column of parakeratotic cells that extend through the surrounding orthokeratotic stratum corneum.
Fig. 1. A single, discrete, firm, 1 mm, seedlike keratotic plug similar in appearance to a music box spine on the left 3rd finger.

Reduced to absent granular layer beneath the column of parakeratosis, and vacuolated or dyskeratotic cells in the underlying epidermis. Cornoid lamellation is a constant feature of many variants of porokeratosis and is an occasional associated finding in other inflammatory, hyperplastic or neoplastic diseases of the skin. But Friedman et al. and Osman et al. insisted that the spiny lesions should not be classified as porokeratosis because they lack the centrifugally expanding rings of porokeratosis and do not coalesce to form plaques. In addition, electron microscopic findings of the epidermis beneath the orokeratotic column show no evidence of dyserkeratosis, vacuolar degeneration or exaggerated lumping of tonofilaments as seen in orokeratosis. They also insisted that “spiny keratoderma” was a more appropriate term than “punctate keratoderma” because clinically punctate keratoderma has hyperkeratotic papules, and histologically, simple hyperkeratosis with underlying acanthosis and minimal parakeratosis.

Eight case reports of “spiny keratoderma” have appeared involving 15 patients (Table 1). Family history showed 4 instances of transmission in a dominant fashion, and males were more often affected than females. It developed after puberty, generally between the second and fourth decades, with age of onset ranging from 12 to 70 years. The spiny lesions were not of sudden onset, but progressed slowly and remained asymptomatic. They were bothersome primarily for cosmetic and aesthetic reasons and had not been reported to resolve spontaneously. Numerous therapeutic attempts at removal have been unsuccessful. Recently Osman et al. reported a good result obtained by using topical 5-fluorouracil cream but the lesion recurred after discontinuance of the treatment. Authors differ about which cases are consistent with "spiny keratoderma." Although Osman et al. insisted it should be confined to the spiny lesions on hands and feet without other cutaneous findings, Sakas and Gentry and Shiff

Fig. 2. A central parakeratotic column, so called cornoid lamella-like, is seen arising a slightly depressed area of the epidermis with a reduced granular layer (H & E stain, A: ×40, B: ×200).
Table 1. Case reports of spiny keratoderma

<table>
<thead>
<tr>
<th>Case</th>
<th>Authors</th>
<th>Age/Sex ; Race</th>
<th>Distribution</th>
<th>Family Hx</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Brown^1</td>
<td>20/M ; white</td>
<td>BPS ; FD</td>
<td>YES</td>
</tr>
<tr>
<td>2</td>
<td>Herman^2</td>
<td>50/M ; white</td>
<td>BPS ; FD</td>
<td>NO</td>
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<tr>
<td>3</td>
<td>Sakas &amp; Gentry^4</td>
<td>60M ; Oriental</td>
<td>BPS ; FD</td>
<td>NO</td>
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<tr>
<td>4</td>
<td>Friedman et al^5</td>
<td>72/M ; - -</td>
<td>BPS ; FD, DD</td>
<td>NO</td>
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<tr>
<td>5</td>
<td>Fridman et al^6</td>
<td>55/F ; - -</td>
<td>BPS ; FD, LD</td>
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</tr>
<tr>
<td>6</td>
<td>Lestringant &amp; Berge^5</td>
<td>75/M ; white</td>
<td>BP ; FD</td>
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<td>7</td>
<td>Lee et al^7</td>
<td>60/f ; Korean</td>
<td>BPS</td>
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<tr>
<td>8</td>
<td>Lee et al^7</td>
<td>66/M ; Korean</td>
<td>BPS</td>
<td>YES</td>
</tr>
<tr>
<td>9</td>
<td>Osman et al^8</td>
<td>72/F ; Hispanic</td>
<td>BPS ; FD, LD</td>
<td>NO</td>
</tr>
<tr>
<td>10</td>
<td>Present Case</td>
<td>62/M ; Oriental</td>
<td>LD</td>
<td>NO</td>
</tr>
</tbody>
</table>

BP : Bilateral Palms ; BPS : Bilateral palms and soles ; DD : dorsal aspects of digits
FD : Flexor aspects of digits ; LD : Lateral aspect of digits

and Hughes^6, included in their series patients with involvement of other cutaneous sites, such as, unilateral and linear distribution as well as patients with facial sebaceous hyperplasia. Like Sakas and Gentry and Schiff and Hughes' cases, although our patient had a single spiny lesion on the lateral aspect of the finger, it might be another variant of "spiny keratoderma" and more cases will be reported showing anothe variant forms of clinical features.

REFERENCES