Sclerotic Fibromas of the Skin
— Reports of Two Cases and a Review of the Literature in Japan —

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Summary: Two cases of sclerotic fibromas of the skin are presented. Clinical and light-microscopic findings are also reviewed from 8 sclerotic fibromas of the skin arising in other Japanese patients. The patients in this series had no manifestations of Cowden's disease.

Key words: sclerotic fibromas of the skin — collagen fiber — Cowden's disease — Japanese — vimentin

Introduction

In 1989, Rapini and Golitz described eleven cases with a peculiar type of dermal fibroma that is almost acellular. They suggested the term, sclerotic fibromas of the skin. Two new cases with sclerotic fibromas of the skin are presented in this report along with a review of the Japanese literature.

Reports of Cases

Case 1: A 21 year-old man came to the Dermatology Clinic at Kurume University Hospital with a solitary tumor on the left forehead. The tumor had been present for at least 1 year, and it was gradually increasing in size. The patient was asymptomatic. A physical examination revealed a solitary 1.5 × 1.5 cm soft, skin-colored tumor that was not fixed to the underlying tissue. There was no lymphadenopathy. The tumor was completely excised. There has been no evidence of a recurrence, 18 months post-resection.

Case 2: A 59 year-old man, apparently healthy and alert, visited the clinic complaining of a papule on the left palm which had been present for approximately thirty years. On physical examination, the papule was 0.9 × 0.9 cm in size, waxy white yellow in color, elevated, oval, firm, and well circumscribed. An apparent connection was present between the papule and the skin surface, however, the papule was not fixed to the underlying tissue. The tumor was removed and at a 14 month follow-up, the patient had no evidence of a local recurrence.

Histopathology: The specimens from the two patients had essentially the same histopathologic appearance. The epidermis was atrophic. The lesions were well-demarcated, non-encapsulated, round or elliptically shaped tumors in the upper and mid-dermis, separated from the epidermis by a narrow grenz zone. The tumors had distinctive tortuous appearances; focally there was a whorl-like pattern. The collagen fibers were finely fibrillar, and strikingly hyalinized like relatively acellular keloids (Fig. 1). Al-
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cian blue staining demonstrated an increased amount of acid mucopolysaccharide within these spaces. Weigert-van Gieson and Masson trichrome stains showed that the hyalinized areas were similar to those of collagen, and elastic tissue was absent or nearly absent within the lesions. The periodic acid-Schiff stain, with or without diastase, accentuated the staining of the hyalinized collagen, as compared to normal surrounding collagen. Staining for S-100 protein, neuron-specific enolase and desmin was negative, however, immunoreactivity for vimentin was observed.

Discussion

Starink et al. (1985) histologically studied 40 cutaneous biopsies from 7 patients with Cowden's disease, and observed that 9 biopsies, 5 of which were taken from pearly facial papules and flat nodules, had a special type of fibroma. The tumor was a distinctive type of fibroma characterized by an organized pattern of interwoven fascicles of collagen bundles with a laminated or tortuous appearance. They suggested that the tumors represented a second microscopic hallmark of Cowden's disease, in addition to the facial trichilemmoma.

Rapini and Golitz (1989) first described the solitary occurrence of the same fibroma in patients without Cowden's disease and called it a sclerotic fibroma of the skin. The lesions were waxy white, translucent, or flesh-colored solitary papules. They ranged in size from 3-9 mm in diameter (mean, 6 mm) and were present for "several months" to eight years. The average age of the patients was 40 years (range, 7-62 years). None of the patients were known to have Cowden's disease. There was no characteristic location for the lesions, which occurred on the face, trunk, and upper and lower extremities. The authors mentioned the possibility that the tumors were simply

Table 1.

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Location</th>
<th>Duration</th>
<th>Maximum diameter (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>39</td>
<td>M</td>
<td>rt inguinal</td>
<td>2 mo</td>
<td>2</td>
</tr>
<tr>
<td>2</td>
<td>49</td>
<td>M</td>
<td>forehead</td>
<td>2 yr</td>
<td>bean</td>
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<tr>
<td>3</td>
<td>78</td>
<td>F</td>
<td>rt 2nd finger</td>
<td>40 yr</td>
<td>16</td>
</tr>
<tr>
<td>4</td>
<td>34</td>
<td>M</td>
<td>lt palm</td>
<td>8 yr</td>
<td>8</td>
</tr>
<tr>
<td>5</td>
<td>51</td>
<td>M</td>
<td>base of nose</td>
<td>4 yr</td>
<td>9</td>
</tr>
<tr>
<td>6</td>
<td>21</td>
<td>M</td>
<td>forehead</td>
<td>1 yr</td>
<td>15</td>
</tr>
<tr>
<td>7</td>
<td>59</td>
<td>M</td>
<td>lt palm</td>
<td>30 yr</td>
<td>9</td>
</tr>
<tr>
<td>8</td>
<td>35</td>
<td>M</td>
<td>rt retroauricle</td>
<td>10 yr</td>
<td>9</td>
</tr>
</tbody>
</table>
SCLEROTIC FIBROMAS OF THE SKIN

a variant of a dermatofibroma. The following features seemed to make them distinct: 1) clinical appearance; 2) atrophic epidermis (uncommon in ordinary fibromas); and 3) hypocellular, hyalinized dermal collagen bands separated by prominent clefts often containing abundant acid mucopolysaccharide.

A review of the Japanese literature disclosed 7 cases with sclerotic fibromas of the skin (Table 1). The lesions were waxy white, skin colored, flesh-colored and slightly brown solitary papules. They ranged in size from 2-16 mm in diameter (mean, 9.8 mm) and were present for 2 months to 40 years. The average age of the patients was 46 years (range, 21-78 years). There was no characteristic location for the lesions. None of the patients were known to have Cowden’s disease. As mentioned above, sclerotic fibromas of the skin occurring in the Japanese have the same clinical features as the fibromas described by Rapini and Golitz (1989). We agree with the concept that the tumor represents an unrecognized form of the same fibrous hamartoma that occurs in Cowden’s disease.

References