increase the risk of developing squamous cell tumours (18, 19). Thus, in spite of its efficacy in recalcitrant PPP, oral CsA should be reserved for disabling forms of PPP in patients who are unresponsive to or intolerant of standard therapies.

REFERENCES


Multiple Familial Eccrine Spiradenoma with Cylindroma

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This report documents an association between multiple eccrine spiradenomas and multiple cylindromas in three generations of one family and provides further evidence to support previous contentions that these tumours are derived from a pluripotential basal cell. We propose that eccrine spiradenoma may be inherited as an autosomal dominant. Key words: Eccrine sweat gland; Basal cell; Autosomal dominant.

(Accepted May 28, 1989.)

Acta Derm Venereol (Stockh) 1990; 70: 79–82.

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Eccrine spiradenoma is almost invariably present as a solitary tumour, most commonly located on the chest and face (1). Multiple eccrine spiradenomas have occurred in individual patients (2) and in association with multiple cylindromas and trichoepitheliomata in three members of one family (3); an association
between multiple eccrine spiradenomata, cylindromata and trichoepitheliomata has been noted in individuals (4, 5). While multiple cylindromata are recognized to be inherited in an autosomal dominant pattern (6), eccrine spiradenomata are stated not to be familial (7).

We present 5 members of one family with multiple tumours that proved histologically to be eccrine spiradenomata and cylindromata and we propose that eccrine spiradenoma too may be dominantly inherited.

CASE REPORT
The first member of this family to present to the Department of Dermatology is shown as Case 1 in Fig. 1. She first developed lumps on the skin during childhood and has continued to develop new lesions over the following six decades. These have occurred most commonly on the trunk and scalp, and vary in size between 3 and 20 mm. Some are tender. Her mother is said to have had clinically identical lumps but resisted surgery until she died at the age of 87 years from unrelated causes.

Case 1 had 5 brothers, of whom 2 survive. None of these had lumps. Of her 4 surviving sisters, 3 have multiple cutaneous tumours (Cases 2, 3 and 4 in Fig. 1). Both children of Case 2 have lumps, although only the daughter (Case 5; Fig 1) has been examined.

Examination of Case 1 repeatedly over 26 years has shown multiple dermal papules and nodules, some with a bluish hue, although most were skin-coloured. There was no visible difference between those which were tender and those which were non-tender. She has had 53 tumours submitted for histological examination over that time.

While the clinical and histological features of these cases is shown in Table 1, attention is drawn to the pattern of tumours in Case 5 (Fig. 2). Multiple tender sub-cutaneous nodules were scattered in a linear, zosteriform pattern over the left mastoid bone.

Fig. 1. Affected family members. ● Female, □ male. Those family members examined by the authors clinically and histologically are marked by an asterisk. The numbers refer to the Case numbers used in the text.

Histology
In Case 5, all tumours examined show the typical histological appearances of eccrine spiradenoma, with a sharply demarcated lobular appearance, and comprising interlacing cords of epithelial cells (Fig. 3). Small lumina are sometimes seen. Hyaline material is present in the stroma of many of the tumours.

In all the other cases described, histological examination has shown some of the tumours to be eccrine spiradenomas, while the others comprise islands of tumour cells of varying size, surrounded by a hyaline sheath, with the typical 'jigsaw' appearance of cylindroma. Tubular lumina are frequently seen. The tumour islands contain cells with deeply staining nuclei arranged in a palisade around the periphery and cells with larger, lighter staining nuclei in the centre (Fig. 4). In several cases, the two distinct tumour types are seen directly abutting each other (Fig. 5).

Fig. 2. Linear arrangement of subcutaneous nodules overlying the left mastoid bone (Case 5).

Fig. 3. Eccrine spiradenoma (Case 1) H & E. ×400.


DISCUSSION

These case reports document the appearance of multiple eccrine spiradenomata and cylindromata in two generations of one family, with reliable reports of similar tumours in a third generation.

The pathogenesis of eccrine spiradenoma is controversial. Kersting & Helwig recognized the sweat gland origin of this tumour and considered it to be derived from the eccrine sweat gland anlage (1). It was a solitary tumour in 97% of their 134 cases, and though multiple in 2 of their cases, in no case was it familial. Munger et al. (8) concluded that the cells composing an eccrine spiradenoma were similar to eccrine sweat gland cells. Further electron microscopic studies of a case of multiple eccrine spiradenomata suggested to the authors that the tumours are composed of basal cells situated on or near a basement membrane, apparently differentiating towards eccrine structures as they moved away from the basement membrane (9).

Castro & Winkelmann (10) cite additional ultrastructural evidence that the eccrine spiradenoma is derived from basal cells. In particular, they found Langerhans’ cells in the spiradenoma; these are also present in cylindromata and basal cell carcinomata (11) as well as in normal stratifying squamous epithelium. On the other hand, Cotton et al. (12) cite recent immunohistochemical studies in support of an eccrine origin for eccrine spiradenomas, in particular the demonstration that spiradenomas express simple keratins characteristic of eccrine epithelium.

The coexistence of cylindroma and eccrine spiradenoma, although extremely rare, is well known. In Crain & Helwig’s review of cylindroma, 2 out of 56 cases had eccrine spiradenomas as well (5). Subsequently scattered case reports have confirmed the

Table I. Characteristics of cutaneous tumours in five members of one family

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Sex</th>
<th>No. of tumours</th>
<th>Size</th>
<th>Site</th>
<th>Age at onset</th>
<th>Pain</th>
<th>Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Trunk and scalp</td>
<td>Childhood</td>
<td>Present</td>
<td>Cylindroma</td>
</tr>
<tr>
<td>1</td>
<td>F</td>
<td>&gt; 53</td>
<td>3–20 mm</td>
<td>Upper back and scalp</td>
<td>20's</td>
<td>Absent</td>
<td>+</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>&gt; 24</td>
<td>2–15 mm</td>
<td>Scalp, face, trunk</td>
<td>30's</td>
<td>Present on scale</td>
<td>+</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>&gt; 20</td>
<td>3–20 mm</td>
<td>Scalp, neck, back</td>
<td>Present in some</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>6</td>
<td>4–15 mm</td>
<td>Scaph, face, zosteriform</td>
<td>Childhood</td>
<td>Marked</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>&gt; 15</td>
<td>2–10 mm</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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coexistence of these tumours (3, 4, 13). Furthermore, features typical of eccrine spiradenoma and cylindroma have existed in the same tumour from one patient (13). The occurrence of a cluster of eccrine spiradenomata in a linear or zosteriform pattern echoes previous reports, but is of unknown significance (14, 15). In none of the previous reports did the spiradenomata occur in association with cylindromata, but this zosteriform pattern must now be recognized as characteristic of multiple eccrine spiradenomata.

The family we report here add to this apparent association between eccrine spiradenoma and cylindroma in several ways. Their occurrence together adds support to the hypothesis that both these tumours are derived from a pluripotential basal cell. The development of monoclonal antibodies against differentiation markers should enable a more precise determination of this relationship to be made. The occurrence of multiple eccrine spiradenoma with multiple cylindromata across three generations of one family is most consistent with an autosomal dominant pattern of inheritance and confirms the conclusion of the single existing case report of a familial incidence that eccrine spiradenoma may be inherited in this way.

REFERENCES

Adrenergic Urticaria and Adrenergic Pruritus

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We report here on 2 patients with adrenergic urticaria and adrenergic pruritus, respectively. The lesions and features developed during phases of stress and during the attacks were associated with an increase in the plasma concentrations of noradrenaline, adrenaline and prolactin. The dopamine plasma level was elevated only in the case of adrenergic urticaria. The symptoms could be reproduced by intradermal injection of adrenaline and noradrenaline and treated successfully with propranolol, a blocker of β-adrenergic receptors. Adrenergic urticaria is a rare but distinct entity, which has to be separated from cholinergic urticaria. Adrenergic pruritus seems to be a minor variant of adrenergic urticaria. Key words: Noradrenaline; Adrenaline; Cholinergic urticaria.

(Received June 7, 1989.)

Acta Derm Venereol (Stockh) 1990; 70: 82–84.

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Eliciting factors of most types of physical urticaria are well-defined (1). In 1985, Shelley & Shelley (2) de-