Multifocal Papillary Apocrine Adenoma Arising in a Systematized Linear Epidermal Nevus

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Previous reports indicate that a linear epidermal nevus (LEN) may serve as a source of development for additional skin tumors. We report an unusual case of multifocal papillary apocrine adenoma (PAA) arising in an LEN. This is the first case of its kind documented in the literature. Cutis. 2004;73:348-352.

Epidermal nevi are a proliferation of keratinocytes and skin appendages of unknown etiology. They arise from pluripotent cells in the germinal layer of the embryonic ectoderm. The most common form of epidermal nevus is a linear epidermal nevus (LEN). An LEN is mostly asymptomatic, and, when extensive, cosmetically disfiguring. Rarely, neoplasms such as basal cell carcinoma, squamous cell carcinoma, and keratoacanthoma can arise in an LEN.1-11 We describe a case of multifocal papillary apocrine adenoma (PAA) arising in an LEN.

Case Report
A 31-year-old otherwise healthy woman was seen in our clinic for a brown-black, rough lesion on her left upper chest. This lesion had been present since birth and had slowly increased in size. It was mostly asymptomatic, with occasional episodes of bleeding after irritation from the patient’s clothing. There was no family history of similar lesions. The physical examination revealed a linear configuration of brown-black, verrucous, cutaneous plaques and papular clusters on the left anterior chest wall (Figure 1). The plaque over the sternum measured $10.5 \times 3.0 \times 0.6$ cm and extended from the second intercostal space to the xiphoid process. The portion over the left upper chest area measured $7.5 \times 3.0 \times 0.6$ cm. Just lateral to these main plaques were a few verrucous clusters that extended up to the anterior axillary fold. Similar tan-yellow plaques on the patient’s right hand (both palmar and dorsal aspects) and tan-brown, minimally keratotic plaques on the left medial forearm were noted. Clinically, the patient was diagnosed to have an LEN, systematized type.

The benign nature of the cutaneous processes was explained to the patient. The patient opted for removal of her chest lesions secondary to irritation and cosmetic reasons. The LEN was excised surgically in 2 stages, with a 6-month interval between excisions. The 4- to 6-week postsurgery follow-up examination revealed slightly hypertrophic scars. The patient subsequently failed to maintain her follow-up appointments.

The resected specimens from both surgeries were sent for histopathologic examination. The excised specimens measured $10.5 \times 3.0 \times 1.5$ cm (midsternal nevus), $7.5 \times 3.0 \times 1.5$ cm (left upper chest nevus), and $1.0 \times 0.8 \times 0.8$ cm (papillary clusters). All 3 specimens showed a rough, black verrucous surface surrounded by a small rim of normal skin. The specimens were formalin fixed, and the sections were stained with hematoxylin-eosin. Histologically, the sections showed features characteristic of an LEN, with hyperkeratosis, acanthosis, and papillomatosis. A focal band of lymphocytic inflammation in the papillary dermis accompanied the epidermal proliferation. The presence of well-formed sebaceous lobules, aborted or abnormal hair follicles, or both, and apocrine glands deep in the dermis (features characteristic of nevus sebaceus) were not observed in any of the

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The authors report no conflict of interest.

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sections. However, the unusual feature was the presence of multiple foci of well-circumscribed nodules of ductular proliferation in the dermis (Figure 2). Each nodule was composed of 4 to 12 dilated tortuous ducts with prominent intraluminal papillary units with occasional bridging (Figure 3). A few of the dilated ducts were filled entirely with multiple papillary units, giving a solid appearance to a few nests. An outer layer of flattened myoepithelial cells and an inner layer of tall columnar cells, showing some pseudostratification (Figure 4), lined the ducts. Features of apocrine differentiation, such as the presence of eosinophilic cytoplasm, decapitation secretions, and apical knobs, were evident in the neoplasm. The nodules did not exhibit any infiltrating growth pattern.

**Comment**

Epidermal nevi are developmental (hamartomatous) malformations of the skin, characterized by hyperplasia of epidermal structures. These nevi may be keratinocytic, follicular, sebaceous, eccrine, or apocrine in origin. The incidence ratio is 1:1000 live births. The most common type of epidermal nevi is keratinocytic nevi, also called linear epidermal nevi. They often appear at birth or infancy and slowly enlarge during childhood. They usually reach a stable size by adolescence. Most LEN occur sporadically, though rare familial occurrences have been described. There is no sex predilection.

Clinically, LEN are characterized by closely set, skin-colored or brown-black verrucous papules often coalescing into well-demarcated plaques. The linear arrangement typically follows the Blaschko lines. LEN may be localized or diffuse. LEN with diffuse or extensive distribution are called systematized epidermal nevi. When located on one half of the body, they are called nevus unius lateris. Ichthyosis hystrix is an epidermal nevus with extensive bilateral distribution and may be associated with developmental abnormalities in other systems (the epidermal nevus syndrome). Inflammatory linear verrucous epidermal nevus is an inflammatory variant of the epidermal nevus, with a characteristic histologic pattern of alternating hyperkeratosis with hypergranulosis and parakeratosis with hypogranulosis.

Different histopathologic patterns of LEN have been described. The most common pattern is a simple squamous papilloma, characterized by hyperkeratosis, acanthosis, and papillomatosis with elongated rete ridges. Occasionally, epidermolytic hyperkeratosis, Darier disease, seborrheic keratosis, verruca, porokeratosis, acanthosis nigricans, and
acrokeratosis verruciformis-like patterns have been described in LEN.\textsuperscript{15}

The development of neoplasms, benign or malignant, epidermal or adnexal, is well documented in nevus sebaceus of Jadassohn.\textsuperscript{16,17} Their incidence ranges from 6.5% to 22% according to various reports. Rarely, however, neoplasms have been described arising in an LEN. A review of the literature revealed 13 such cases,\textsuperscript{1-13} which are summarized in the Table. Except for one case\textsuperscript{13} of adnexal origin, all other tumors were of epidermal origin.\textsuperscript{1,12} Most neoplasms were diagnosed in early-to-mid-adult life, long after the initial appearance of the LEN.

The LEN we describe in this case report is very unusual. Although the nevus had microscopic features characteristic of an LEN, a unique and unusual finding was the presence of a multifocal PAA in the dermis. Despite the lesion being multifocal, we believe that the apocrine glandular proliferation present in the dermis represents a neoplastic process rather than a hamartoma. To our knowledge, the occurrence of multifocal PAA in an LEN has not been described previously in the literature.

Patients with LEN or their variants usually seek medical care either for relief of symptoms or for cosmetic concerns. Many reports in the literature...
**Review of Different Neoplasms Arising in a Linear Epidermal Nevus**

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age, y (sex)</th>
<th>Location</th>
<th>Diagnosis</th>
<th>Associated Neoplasm(s)</th>
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<tr>
<td>Toyama et al, 1936$^1$</td>
<td>47 (F)</td>
<td>Generalized</td>
<td>Verrucous nevi</td>
<td>BCC, Bowen, SCC</td>
</tr>
<tr>
<td>Pack et al, 1941$^2$</td>
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<td>Right trunk/upper limb</td>
<td>Nevus unius lateris</td>
<td>BCC</td>
</tr>
<tr>
<td>Carney, 1952$^3$</td>
<td>Unknown</td>
<td>Unknown</td>
<td>Epidermal nevus</td>
<td>BCC</td>
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<td>Litzow et al, 1961$^4$</td>
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<td>BCC, multiple</td>
</tr>
<tr>
<td>Swint et al, 1970$^5$</td>
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</tr>
<tr>
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<tr>
<td>Goldberg, 1980$^7$</td>
<td>57 (M)</td>
<td>Left preauricular</td>
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<td>Horn et al, 1981$^8$</td>
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<td>Abdomen</td>
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<td>Cramer et al, 1982$^9$</td>
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<td>Rosen, 1982$^{10}$</td>
<td>32 (M)</td>
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<td>LEN</td>
<td>KA</td>
</tr>
<tr>
<td>Braunstein et al, 1982$^{11}$</td>
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<tr>
<td>Ichikawa et al, 1996$^{12}$</td>
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<td>Trunk</td>
<td>VEN</td>
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<tr>
<td>Hamanaka et al, 1996$^{13}$</td>
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<td>Right arm</td>
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<td>Malignant eccrine poroma</td>
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<td>Present case, 2004</td>
<td>31 (F)</td>
<td>Left chest</td>
<td>LEN, systematized</td>
<td>Papillary apocrine adenoma</td>
</tr>
</tbody>
</table>

*F indicates female; M, male; BCC, basal cell carcinoma; SCC, squamous cell carcinoma; LEN, linear epidermal nevus; KA, keratoacanthoma; and VEN, verrucous epidermal nevus.

**Figure 4.** Tall columnar cells with eosinophilic cytoplasm with terminal knobs and decapitation secretions. Note the layer of myoepithelial cells at the periphery of each duct (H&E, original magnification ×400).
suggest benefit from a variety of treatment modalities, ranging from topical therapy to surgical procedures. However, a full-thickness surgical excision is considered by many to be a very effective form of treatment.

**Conclusion**

In this article, we describe a case of a multifocal PAA arising in an LEN. To our knowledge, this is the first one to be reported in the literature. A full-thickness surgical excision with clear margins appears to be an effective treatment for these types of lesions.

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**REFERENCES**