Blaschkoid Eccrine Spiradenomas

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We report a case of multiple eccrine spiradenomas that arose in adulthood along a patient's arm and continued to the midline of the patient's chest across multiple dermatomes. This is one of a handful of such cases, which have been reported as linear, nevoid, or zosteriform eccrine spiradenomas. We believe this distribution is best described as Blaschkoid rather than zosteriform or linear because these lesions follow the lines of Blaschko rather than those of a single dermatome.

Eccrine spiradenoma is usually a painful solitary gray-pink nodule less than 1 cm in diameter that is sometimes associated with cylindromas. These lesions arise on the head, neck, trunk, or, less commonly, extremities of adults; however, they also have occurred in infancy and at birth. Variants include satellite, multiple, giant, linear, and zosteriform types.[1]

Case Report

A 46-year-old white man presented with a 20-year history of painful nodules. The initial lesion appeared on his right arm, with subsequent involvement of his right chest and forearm. The lesions enlarged slowly and were periodically painful, both spontaneously and to deep pressure. The patient had undergone numerous excisions of the more painful nodules. He was otherwise in good health, and there was no family history of similar lesions.

Physical examination revealed multiple 1- to 2-cm nodules and linear scars in a curvilinear array over the right chest and extending linearly down the right arm. The nodules were firm and mobile, and either flesh colored or purple-blue (Figures 1 and 2). The remainder of the examination was unremarkable. The differential diagnosis included the following painful tumors: angiolipomas, eccrine spiradenomas, leiomyomas, and blue rubber bleb nevi. An excisional biopsy of a nodule was obtained (Figure 3).

Results of a histopathologic examination revealed a circumscribed, sharply delineated nodule in the upper dermis. A small satellite lobule was present. The nodule was composed of small dark epithelial cells with scanty cytoplasm and hyperchromatic nuclei, which surrounded cords of larger pale cells and connective tissue. The cells assumed a trabecular arrangement, and blood vessels were prominent. This pathology was diagnostic of eccrine spiradenomas.

Comment

Eccrine spiradenoma is a benign adnexal tumor that typically appears as a small and firm bluish nodule and is usually tender on palpation. While most present as solitary lesions on the anterior surface of the upper body, cases of multiple tumors exist. Random multiple tumors have been reported to occur on the chest, upper extremities, forehead, and scalp.[2] Linear and zosteriform distributions also have been reported.[2-4] Noto et al[5] described a case of multiple "nevoid" spiradenomas in which the lesions were confined to the right half of the patient's body, from the face to lower limb. It appears that these lesions and the ones in our case followed Blaschko lines because multiple dermatomes were involved. Indeed, based on current concepts, these patterned cases all may be classified as having a Blaschkoid distribution probably arising from a single aberrant cellular clone present during embryogenesis. Other adnexal tumors, including syringomas, eccrine poromas, and syringocystadenoma, have been reported as following Blaschko lines.[6] To our knowledge, eccrine spiradenomas along Blaschko lines have been reported twice, and our case represents a third example.[7,8]

Onset of nodules in the patterned cases may occur at birth; however, they also have occurred later in life, as reported in our case. Familial cases of eccrine spiradenoma have been reported. This is usually seen in the setting of multiple cylindromas and trichoepitheliomas, the Brooke-Spiegler syndrome. Thus, an autosomal dominant inheritance pattern has been proposed.[9]

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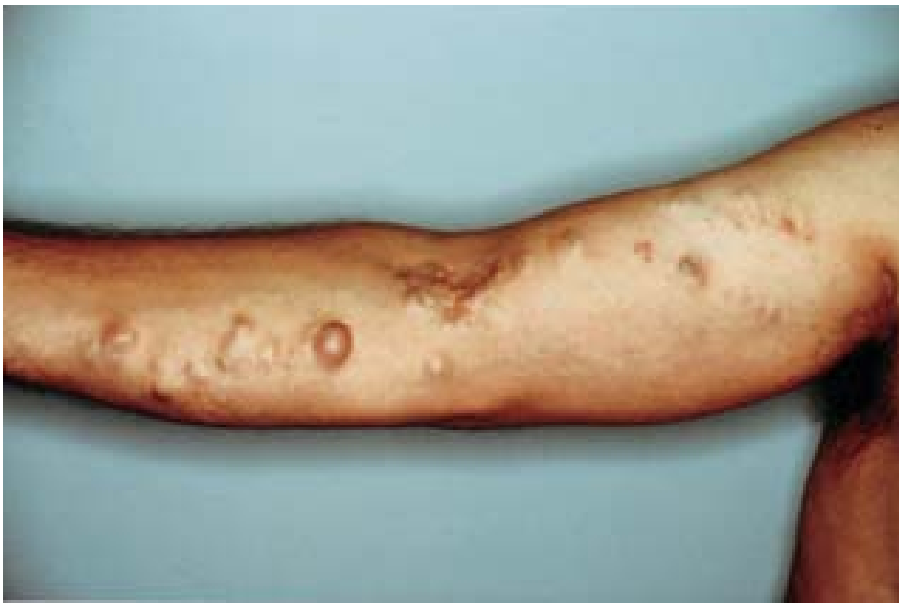

Figure 1. Purple eccrine spiradenoma nodules in a curvilinear array on the medial lateral aspect of the right arm.

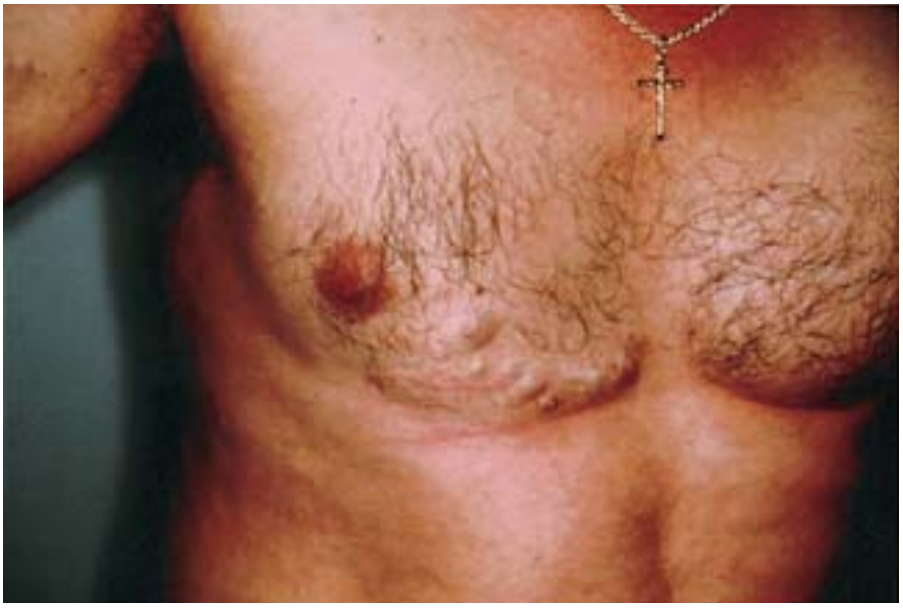

Figure 2. Purple eccrine spiradenoma nodules on the right chest in a curvilinear array stopping at the midline.

The histopathologic hallmark of eccrine spiradenoma is well-demarcated lobules composed of 2 types of cells. Immunohistochemical analysis of these cells has shown that the large pale cells have an immunophenotype that is similar to luminal cells in the transitional portion between the secretory segments and the coiled ducts, and that the small dark cells are similar to those of basal cells in the transitional portion.[10]

Malignant eccrine spiradenoma is a relatively rare phenomenon, with approximately 30 reported cases. These tumors commonly arise within a preexisting benign spiradenoma; malignant change is heralded by rapid growth, change in color, or increased pain.[11] Malignant eccrine spiradenomas have a metastasis rate of greater than 50% in reported cases, with a high resultant mortality rate. It is unclear whether any of these reported malignant cases arose in patients with tumors in Blaschkoid distributions.

Conclusion

Although eccrine spiradenomas are usually solitary and small, our case underscores that a variety of presenta-

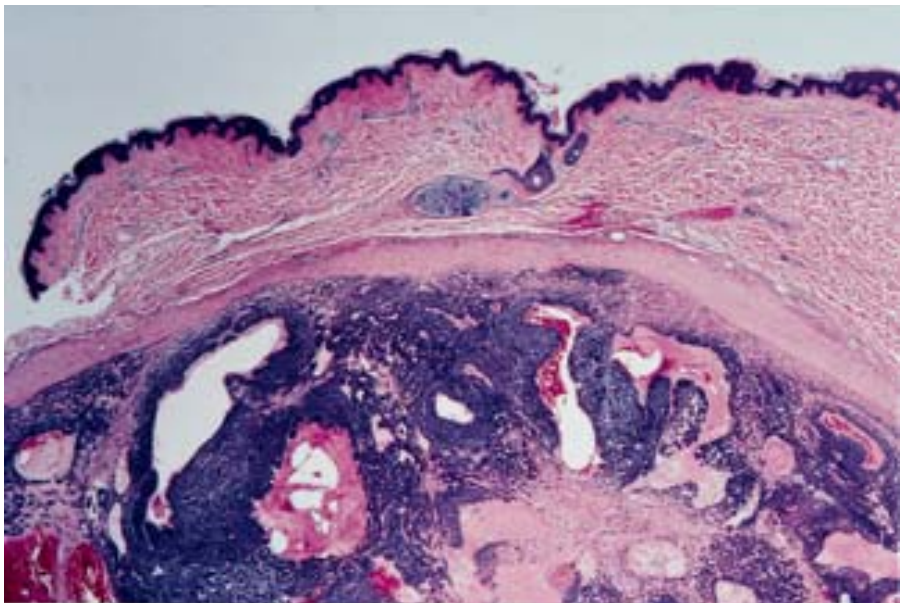

Figure 3. A circumscribed, sharply delineated nodule and small satellite lobule in the upper dermis, composed of small dark epithelial cells with scanty cytoplasm and hyperchromatic nuclei surrounding cords of larger pale cells and connective tissue (H&E, original magnification ×100).

tions are possible. This case also shows that eccrine spiradenoma is another dermatologic condition that can be distributed along the lines of Blaschko. The etiology of this condition, which is undoubtedly genetic, needs further investigation. Why such a Blaschkoid condition arises in adulthood is not well understood and therefore requires exploration.

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