Spiradenocarcinoma of the Scalp

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Spiradenocarcinoma is an exceedingly rare malignant neoplasm with ductal differentiation. Many consider it to be an eccrine neoplasm, and others favor apocrine differentiation. In nearly all cases, spiradenocarcinoma is associated with a benign spiradenoma, with most lesions appearing on the trunk and extremities. We describe a patient who presented with a long-standing and previously asymptomatic scalp nodule that became tender and enlarged. After diagnosis of spiradenocarcinoma, the patient was referred for lymphoscintigraphy, sentinal lymph node biopsy, and Mohs micrographic surgery.

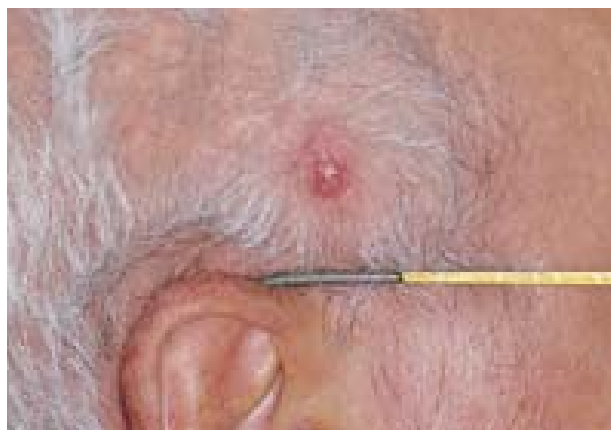

Figure 1. Irregular, firm, tender, nodule of right parietal scalp.

Spiradenocarcinoma (also known as malignant spiradenoma and sweat gland carcinoma ex eccrine spiradenoma)[1] was first described by Dabska[2] in 1972, 16 years after Kersting and Helwig[3] originally described its benign precursor. Lesions are slow growing and usually are several centimeters in diameter, reportedly ranging from 2 mm to 12 cm.[4,5] Most spiradenocarcinomas develop via "malignant degeneration" in long-standing spiradenomas.[6,7] Men and women are affected equally, and most patients with spiradenocarcinoma are older than 50 years.[5,6] Local recurrences, as well as lymph node and distant metastases can occur frequently. Most lesions arise on the extremities and present as chronic solitary nodules that have enlarged recently and become tender or painful.

Histologic diagnosis is made after performing a biopsy on a changing lesion. Sections exhibit signs of malignancy, usually in conjunction with areas of spiradenoma. Wide surgical excision remains the treatment of choice for spiradenocarcinoma.

Case Report

A 65-year-old white man presented with a recently changed scalp nodule. He reported that the lesion had been present for 30 to 40 years and had recently become slightly enlarged, tender, and painful. Examination of the right parietal scalp revealed an approximately 2-cm lobular nodule that was irregular, firm, and tender (Figure 1). The lesion had an opalescent hue with telangiectases and had some resemblance to a basal cell carcinoma (Figure 2). No adenopathy was noted in the head and neck region.

A shave biopsy of the nodule was performed, and sections were stained with hematoxylin and eosin. At scanning magnification (Figure 3), tumor cells were arranged both in a small, well-circumscribed nodule and in variably sized, poorly formed nodules. Higher magnification of the well-circumscribed nodule (Figure 4) revealed homogeneous, eosinophilic material in a reticular pattern, scattered lymphocytes, and 2 distinct types of epithelial cells, namely small and large. The small cells had scant, clear cytoplasm and small, round monomorphous nuclei with condensed chromatin and were situated at the edges of the trabeculae surrounding the large cells. The large cells, that focally form ducts, had features typical of spiradenoma: conspicuous, pale-pink cytoplasm; large nuclei with open chromatin; and occasional small, round nucleoli.

The variably sized nodules were caricatures of the former and extended broadly to the base of the biopsy specimen (Figure 3). Their margins were convoluted, cerebriform, and punctuated by increased

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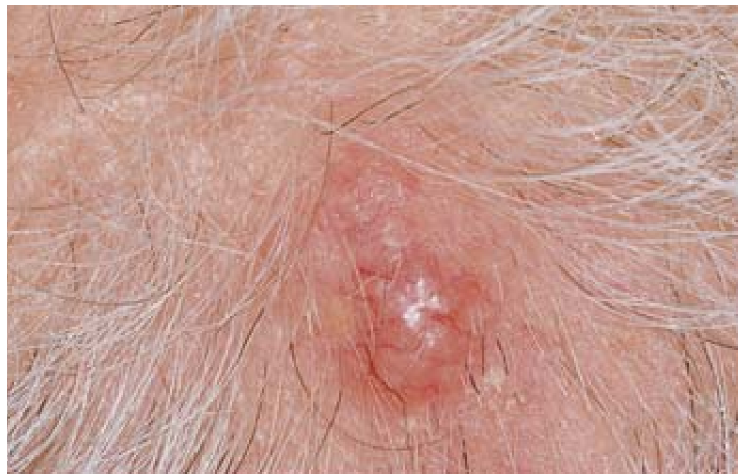

Figure 2. Close-up view of nodule with opalescent hue and telangiectases.

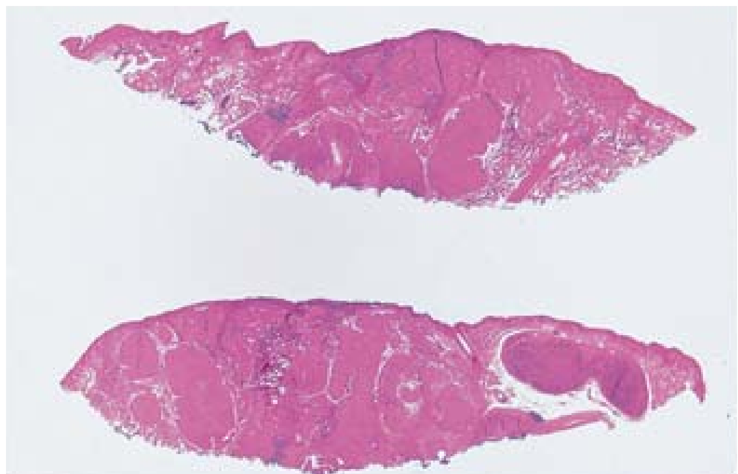

Figure 3. Scanning magnification shows tumor cells in both well-circumscribed and poorly formed nodules (H&E, original magnification ×2.5).

mucin and clefts at the interface with the dermis. Although some nodules had no discernible homogeneous eosinophilic material, others were dominated by it. Higher magnification (Figures 5 and 6) revealed a compelling predominance of large cells with striking cytological features: marked variability in size, bubbly cytoplasm, large nuclei with irregular profiles, vesicular chromatin, prominent and irregularly shaped nucleoli, single-cell necrosis, and numerous mitotic figures. Together, the findings were consistent with spiradenocarcinoma in conjunction with a spiradenoma.

The patient underwent lymphoscintigraphy in anticipation of possible sentinel lymph node biopsy. Three lymph nodes were identified around the right ear and one at the angle of the jaw. Mohs micrographic surgery was performed, which cleared the tumor in one stage. The primary lesion was approximately 14×16 mm. No lymph node dissection was performed once the lesion was revealed to be less than 2 cm. Baseline chest x-ray and liver function test results were within normal limits.

Comment

We describe a case of spiradenocarcinoma of the scalp. To our knowledge, only 2 cases of spiradenocarcinoma in this location have been reported in the literature,[5,8] and, in all, less than 40 cases of this unusual neoplasm have been described. These tumors appear predominantly on the upper extremities, especially the hand.[9] Diagnosis usually is made after a biopsy is performed on a long-standing lesion that becomes acutely painful, tender, or enlarged. Fine needle aspiration has been reported to be an effective, noninvasive diagnostic technique[10] but is not standard practice in the diagnosis of this neoplasm.

Histologic examination revealed areas of spiradenoma, with either gradual or abrupt transition to a malignant growth.[11] Features of the malignant growth included large epithelioid cells, nuclei with open chromatin, and conspicuous nucleoli. Increased mitoses and pleomorphism also were present.[11,12] Immunohistochemical analysis was investigated but appeared to be of little value in distinguishing spiradenocarcinoma from spiradenoma.[7,13]

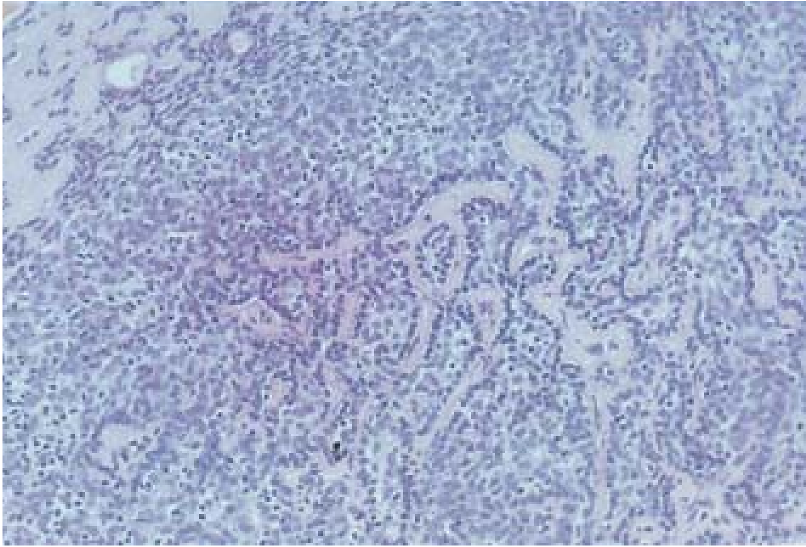

Figure 4. Higher magnification of well-circumscribed nodule shows 2 distinct cell types with scattered lymphocytes (H&E, original magnification ×200).

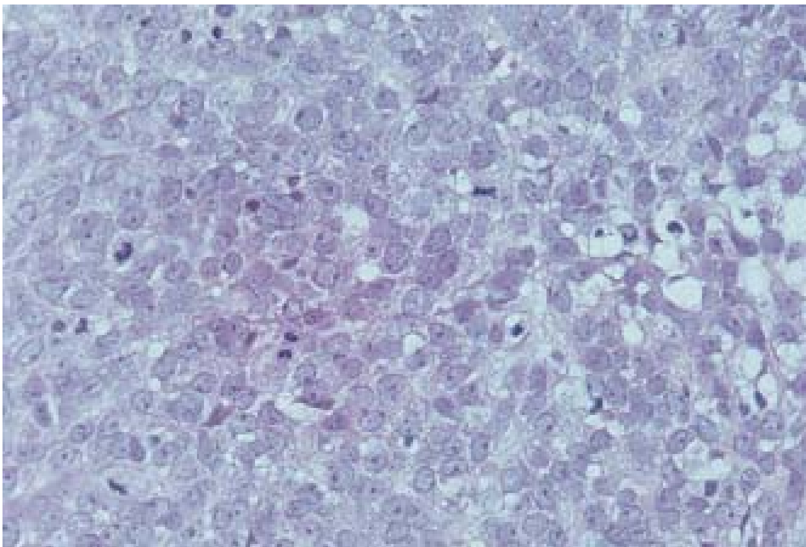

Figure 5. High magnification of poorly formed nodule shows crowded cells with large, irregular, vesicular nuclei, numerous mitoses, and bubbly cytoplasm (H&E, original magnification ×400).

Spiradenocarcinomas have been found in conjunction with cylindromas and trichoepitheliomas,[13-15] and have been reported with rhabdomyoblastic change[6] and foci of osteosarcoma.[6,15,16] Bowenoid features,[15] squamous differentiation,[11,15] and carcinomatous and sarcomatous elements[12,17,18] also have been seen. To our knowledge, one case of malignant chondroid syringoma in a benign spiradenoma has been reported,[19] and 2 separate malignant eccrine spiradenomas were reported in a patient with multiple benign eccrine spiradenomas.[12] Metastases have occurred in nearly 40% of cases and local recurrences in up to 57%.[20] Distant spread to the liver, lungs, brain, bone, skin, and spinal cord has been documented.[4,6]

Surgical excision is required in all cases of spiradenocarcinoma. Complete excision of smaller tumors can be curative. Radiation therapy,[20] chemotherapy,[20] hormonal manipulation,[21] and hyperthermic limb perfusion chemotherapy[20] have been attempted for unresectable and metastatic disease, with little success.

Clinical adenopathy justifies regional lymph node analysis. Lymph node metastasis generally infers a poor prognosis.[20] The role of elective lymph node dissection in detecting asymptomatic lymph node metastases has not been proven to be of therapeutic value. Our patient was referred for lymphoscintigraphy to evaluate for possible sentinal lymph node biopsy. Four lymph nodes were identified around the right ear and jaw. The limited amount of literature on this tumor entity suggests a lower risk for metastasis in lesions less than 2 cm.[6] Because of the size of the lesion, we elected to treat the tumor

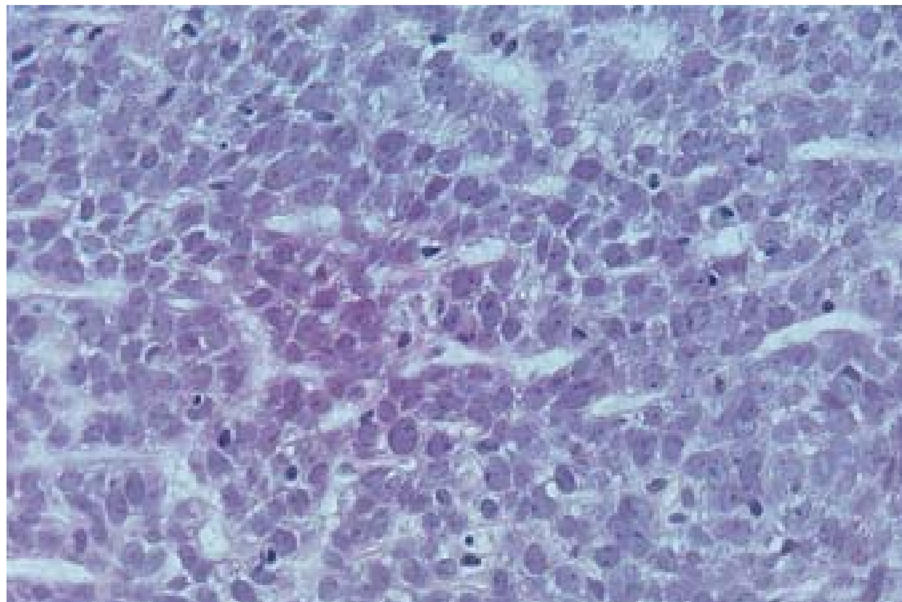

Figure 6. High magnification of poorly formed nodule shows large cells containing variably sized nuclei, irregularly shaped nucleoli and tubules (H&E, original magnification ×400).

with Mohs micrographic surgery and continue to closely monitor the patient clinically. The patient showed no signs of metastasis or recurrence 12 months after diagnosis.

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