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Malignant eccrine spiradenoma: a case report

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Abstract

Malignant eccrine spiradenoma (MES; spiradenocarcinoma) is an extremely rare tumor, which almost always arises from a preexisting eccrine spiradenoma (a benign sweat gland tumor that commonly affects young adults) [1]. We describe a case of malignant eccrine spiradenoma that arose in the right lateral thigh of a 62-year-old woman. The mass, which was present for approximately 35 years, suddenly enlarged with ulceration of the overlying skin. Microscopically, the tumor showed the typical features of an eccrine spiradenoma with areas of adenocarcinoma, squamous differentiation and sarcoma. The tumour qualified for the designation carcinosarcoma arising in eccrine spiradenoma.

Introduction

Eccrine spiradenoma is a benign sweat gland tumor arising from the intradermal straight part of the duct of eccrine sweat glands [1]. The presentation is often that of a single nodule that may or may not be tender. Although benign forms are well recognized, malignant eccrine spiradenoma (MES) is extremely rare and is usually a carcinoma [2,3,4]. Rare cases can have a carcinosarcomatous pattern [5,6,7]. In this report, we present an additional case of MES with carcinosarcomatous pattern and review the literature.

Case Report

A 62-year-old woman presented with a 3 month history of a progressively increasing painless nodule in her right lateral aspect of thigh, which had assumed the form of a large nodular lesion with an ulcerated surface. A small pea sized nodule had

been present since approximately 30 years and for the past 4 months, the lesion had abruptly started to enlarge, with recent ulceration and bleeding. There were no palpable inguinal lymph nodes.

An incisional biopsy was performed which showed predominantly anaplastic spindle cells with frequent mitoses and areas of haemorrhage and necrosis. The lesion was reticulin rich with few reticulin poor clusters. It was reported as malignant neoplasm NOS, and excision was advised.

Subsequently the excision specimen was received. The specimen with overlying skin ellipse, measuring 6.2x 4.5x 3.7 cm, was received in the surgical pathology laboratory. The skin surface showed a nodulo- ulcerative, exophytic, polypoidal growth. The overlying skin was stretched out and showed an ulcer 1.5 cm diameter, covered with exudate. On serial sectioning, a partially encapsulated, multiloculated, cystic and solid, tan-yellow mass measuring 3x3.5x 3.5 cm was noted. The lesion grossly involved the deep dermis and the subcutaneous tissue. However, the margins of resection including the deep resection plane were not involved.

On microscopic examination, the dermis and subcutaneous tissue showed a tumor arranged in ragged sheets, nests, cords, and solid masses with central necrosis along with occasional irregular glandular structures (**Figure 1**). Prominent areas with pleomorphic spindle cells in palisades were observed along with few small foci with squamous differentiation. Areas of tumor necrosis, cystic degeneration, and old hemorrhage were present. The tumor cells had pleomorphic, vesicular nuclei with variably prominent nucleoli and moderate amount of cytoplasm. The mitotic activity was high (both in carcinomatous and sarcomatous areas), however, no lymphovascular invasion was recognized. The overlying epidermis was ulcerated, but did not show any dysplasia. Adjoining to this high-grade carcino-sarcoma, benign eccrine spiradenoma, was seen as sharply demarcated lobules composed of 2 cell populations (peripheral basaloid cells and central pale cells) arranged in nests and inter- anastomosing trabeculae with scattered glandular lumina and focal cystic changes (**Figure 2**). The spindle cell areas were reticulin rich in contrast to the carcinomatous areas, which were reticulin poor (**Figure 3**). Immunohistochemically the sarcomatous areas were positive for Vimentin and negative for smooth muscle Actin (SMA) and Desmin. The carcinomatous areas were negative for Vimentin.

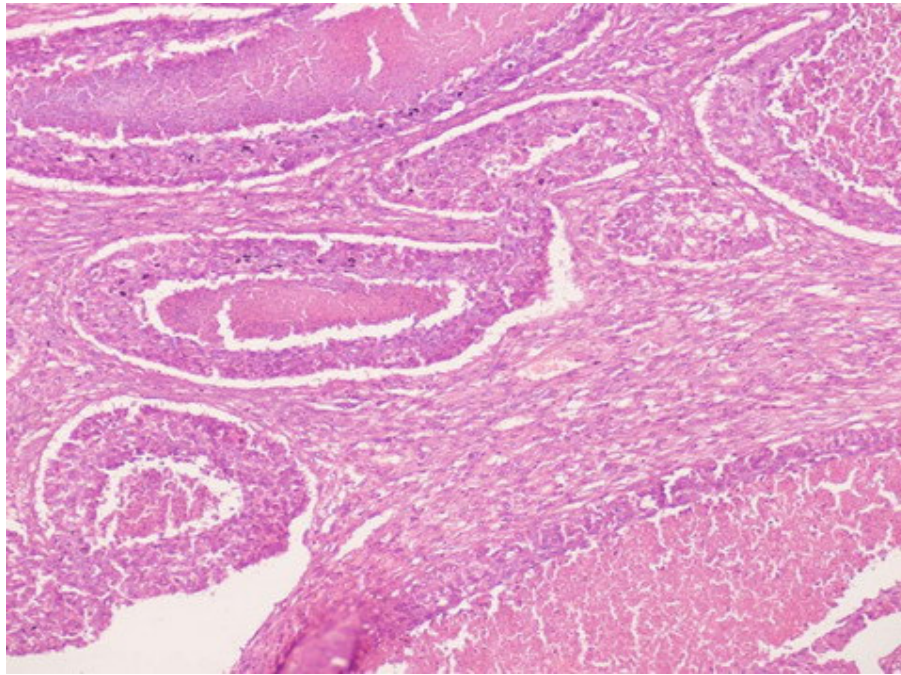


Fig 1: Photomicrograph showing carcinoma with central necrosis and adjacent area showing sarcomatous change (H&E x 100).

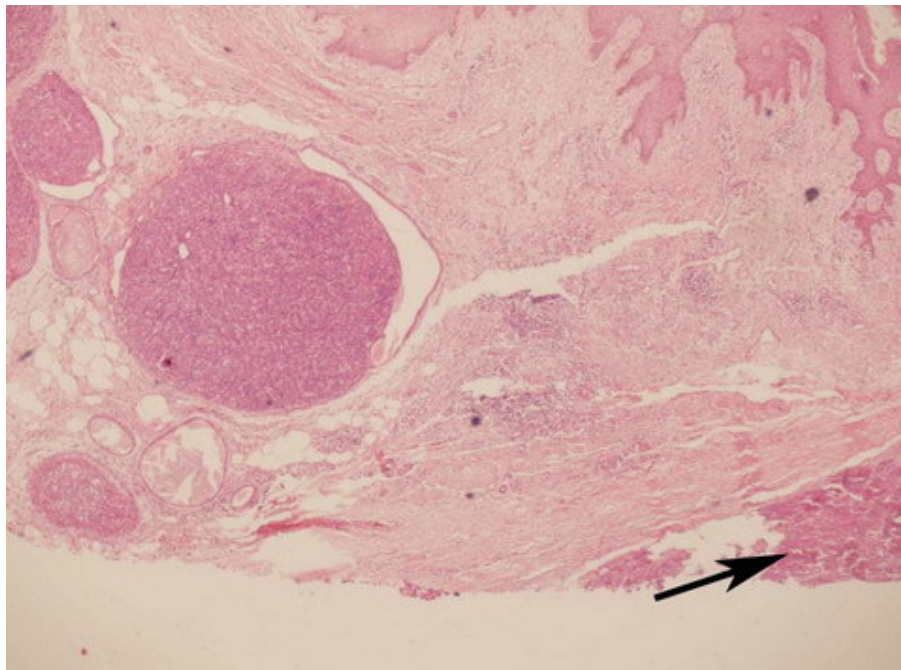


Fig 2: Photomicrograph showing sharply demarcated lobules of benign Eccrine Spiradenoma with a malignant focus shown by pointer (H&E x 40).

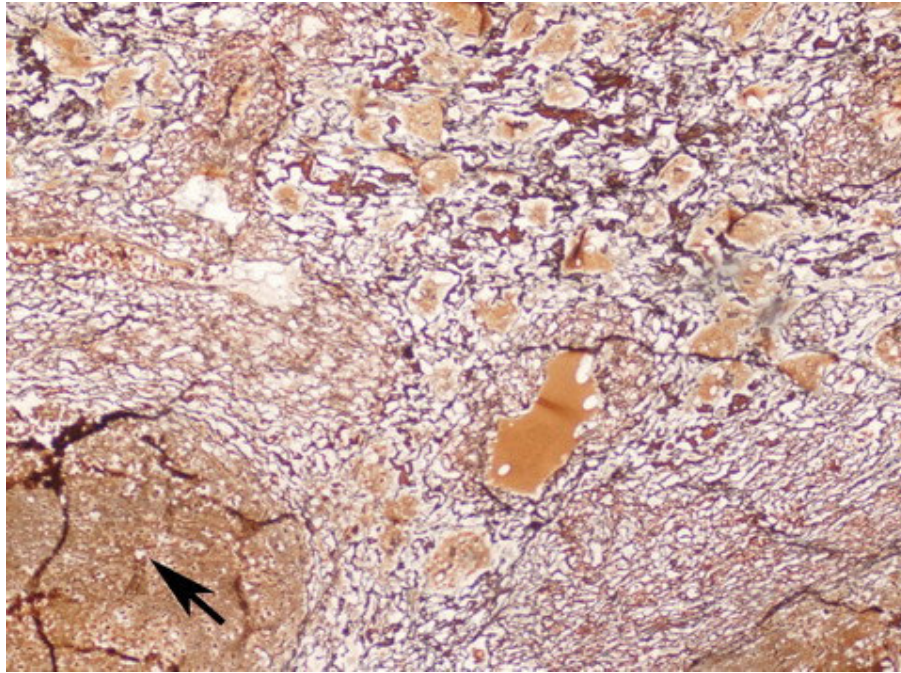


Fig 3: Photomicrograph showing a focus of carcinoma shown by pointer surrounded by reticulin rich sarcomatous areas with prominent vascularity (Reticulin x 100).

Metastatic workup (bone scan, computed tomographic scans of the chest, abdomen, and pelvis) was negative.

Discussion

The motive behind presenting this case was to highlight that malignant transformation in eccrine spiradenoma can have the appearance of carcinosarcoma, with ulceration of the overlying epidermis. The malignant eccrine spiradenoma almost always arises from a pre-existing benign eccrine spiradenoma after a variable latent period, which may be as long as 75 years.[8] It generally begets medical attention when a pre-existing undiagnosed lesion rapidly enlarges, changes color, ulcerates, or becomes painful and tender. This malignant transformation is, fortunately, extremely rare. According to the estimates of Marena and Otto[9], malignant sweat gland tumors account for only 0.005% of all skin tumors.

Clinically, MES presents at an average age of 59 years (range, 21- 92 years) and shows no sex predilection. The tumor usually presents as a solitary firm round dermal nodule on any part of the body but most frequently on the face, scalp, trunk and proximal parts of limbs (92% of reported cases)[2]. Infrequently multiple lesions are present which may become confluent or remain discrete. The overlying epidermis may show no change in color or may be pinkish and rarely ulcerated, as was seen in the present case. The average size of MES at presentation is 3.9 cm (range, 0.5-15 cm)

[2]. Diagnosis of MES is based on histopathologic examination and requires finding a focus of benign spiradenoma within or adjacent to the malignant tumor.[2] Histologically, proliferation of cells with hyperchromatic nuclei, increased mitoses, loss of Periodic-Acid-Schiff positive basement membrane, and invasion of the surrounding tissues characterize malignant transformation in eccrine spiradenoma. MES frequently shows focal squamous differentiation, which may be florid in rare instances.[8] The malignant transformation is usually into a carcinoma; however, carcinosarcomatous transformation has also been reported, as seen in our case. The mesenchymal elements are often nonspecific spindle cells, but rhabdomyosarcoma, osteosarcoma, leiomyosarcoma, and chondrosarcoma have been described in association with carcinoma arising in eccrine spiradenoma.[10,7,5,8,3]

In the absence of a benign focus, the tumor with carcino-sarcoma pattern can be confused microscopically with other malignancies such as squamous cell carcinoma, synovial sarcoma, epithelioid sarcoma and metastatic carcinomas. MES may also be highly vascular and increased vascularity may also lead to diagnostic confusion with a vascular neoplasm.[11]

Immunohistochemically, these tumors exhibit variable expression of cytokeratins, carcinoembryonic antigen, epithelial membrane antigen, and S100 protein.[11,5,8,3] Overexpression of p53 protein in MES has been associated with malignant transformation.[8] Recently these tumors have been shown to demonstrate estrogen receptor positivity, and hormonal receptor status should be evaluated for potential therapeutic options.[11]

Malignant eccrine spiradenoma metastasizes to regional lymph nodes, lungs, brain, and liver (in a descending order of frequency) [2]. While distant metastases of MES are uncommon, they generally portend an ominous prognosis. Appropriate therapy of malignant eccrine spiradenoma consists of a wide local excision with resection of clinically suspicious lymph nodes. Irradiation of the resection site can be useful in preventing local recurrence. The role of chemotherapy is not yet clearly defined. Symptomatic improvement and shrinkage of the tumor with tamoxifen therapy in a patient with estrogen receptor-positive eccrine adenocarcinoma has also been reported [2]. However, the roles of hormonal therapy and other modalities, such as localized postoperative radiation therapy, prophylactic lymph node dissection and chemotherapy still remain to be determined. Close follow-up of these patients for early detection of recurrence and metastases is recommended.

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