

Syringomatous Eccrine Carcinoma of the Vulva

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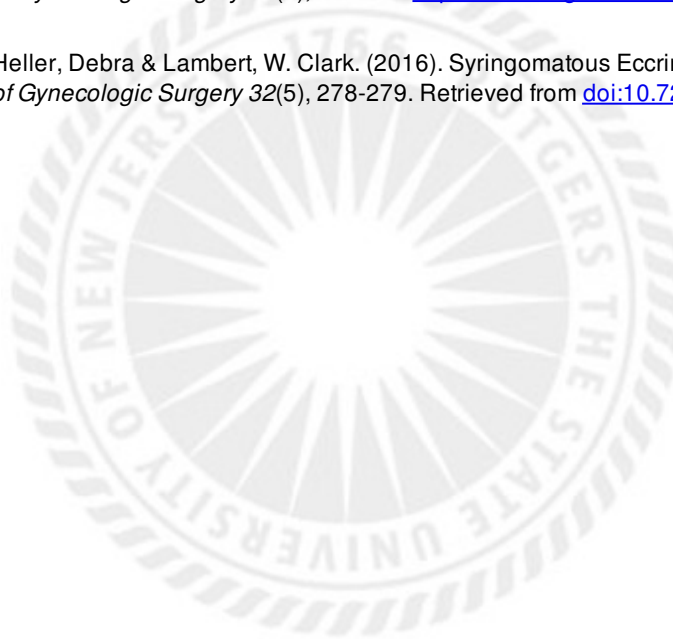
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Syringomatous Eccrine Carcinoma of the Vulva

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Precis:

An exceedingly rare syringomatous eccrine carcinoma arising in the perineum is presented.

Keywords: Vulva, perineum, eccrine carcinoma, sweat gland neoplasms

Abstract

Background: Skin adnexal neoplasms of the vulva are uncommon, and malignant adnexal neoplasms of the vulva are rare.

Case: A case of syringomatous eccrine carcinoma arising in the perineum is presented, and the literature reviewed.

Conclusions: Non-squamous malignancy of the vulva should be considered. All suspicious lesions of the vulva should be biopsied.

Introduction:

The majority of malignancies arising on the vulva are squamous cell carcinomas. Adenocarcinomas rarely arise, and can be classified as extra-mammary Paget disease, Bartholin gland adenocarcinoma, adenocarcinoma arising in anogenital mammary-like glands, or adenocarcinoma of sweat glands(1). Adenocarcinoma arising in vulvar sweat glands is rare(2), and can be apocrine(1), or eccrine. Syringomatous eccrine carcinoma is a rare subtype, with only about 100 cases reported in all body locations(2). It is exceptionally rare on the vulva. A case is reported and the literature reviewed.

Case History:

A woman in her mid-70s presented with a perineal mass. After biopsy confirmation of the diagnosis, she underwent wide local excision of a 6.0 x 5.0 x 3.0 cm solid mass. The lesion came within 2 mm of the resection margin, and showed lymphovascular space involvement, and so the patient was subsequently referred for radiation therapy.

Pathology: The multinodular yellow-tan lesion was seen to erode through the surface epithelium(fig 1). Histologically it was composed of nests and sheets of poorly differentiated epithelial cells(fig 2). At the periphery, a spray pattern of keratinizing infiltrating eccrine ducts(fig 3) confirmed the diagnosis of syringomatous eccrine carcinoma.

Discussion:

Skin adnexal neoplasms of the vulva are uncommon, and malignant adnexal neoplasms of the vulva are rare(2). Classification of malignant sweat gland neoplasms is somewhat confusing, but to some degree parallels the benign counterparts. Hence this tumor, syringoid eccrine carcinoma(SEC), parallels the benign syringoma. SEC is also known by a variety of other names, including eccrine carcinoma and syringomatous carcinoma, among others. Benign syringomas occasionally occur on the vulva, although more often on the face, and present as multiple pruritic flesh colored papules. SEC resembles syringoma in the formation of comma-shaped ductal structures, but also has features of malignancy, including cellularity and invasiveness(2). SEC was first described in 1969(3), and this rare neoplasm is reported predominantly on the scalp, head, neck and trunk(4,5), with only one other case on the vulva reported(2). The lesions do not have a characteristic or consistent immunohistochemical profile(5). SECs are locally aggressive, with a tendency to recur(2,4), and lymph node involvement and distant metastatic disease occur occasionally(4,5). There are no well-established protocols for this rare lesion, the primary treatment is excision. The role of lymph node dissection is controversial, with recommendations including regional node dissection for clinically positive nodes for vulvar sweat gland carcinomas(6), and ipsilateral node dissection for vulvar sweat gland carcinoma, without specification of SEC(7). Of interest is a report of eccrine carcinoma of the vulva associated with extramammary Paget disease. Although not called syringomatous, one of the images does have features suggestive of this diagnosis(8).

In our case, the proximity of the tumor to the margin and lymphovascular space involvement led to the recommendation for adjuvant radiotherapy.

In summary a rare case of vulvar syringomatous eccrine carcinoma is presented. The potential for non-squamous malignancy of the vulva should be considered, and liberal biopsy of all lesions of the vulva performed.

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Disclosures: none

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Legends:

Figure 1-Excision specimen showing tan-yellow lesion ulcerating through skin

Figure 2-Much of the tumor was composed of sheets and nests of poorly differentiated cells

Figure 3-At the edge of the lesion, there was a spray-like pattern of infiltrating keratinizing syringomatous ducts.