

Apocrine adenocarcinoma of the vulva

Babita Kajal, Hetal Talati, Dean Daya, Salem Alowami

Department of Pathology and Molecular Medicine, Mc Master University, Hamilton, Ontario, Canada

Abstract

Cutaneous vulvar carcinomas are predominantly of squamous cell carcinoma type. Primary vulvar adenocarcinomas are rare with a poorly understood histogenesis. They are classified into extramammary Paget's disease, sweat gland carcinomas, and breast-like adenocarcinomas of the vulva. Adenocarcinomas, originating from Bartholin glands, can also present as vulvar adenocarcinoma. Rare adenocarcinomas with apocrine features have been described in the literature. The origin of these neoplasms from the native apocrine sweat glands or from anogenital mammary-like glands is still debatable. We report herein a case of a 67 year old female with a rare primary apocrine carcinoma of the vulva.

Case Report

A 67 year old female presented with an enlarging left vulvar lesion thought to be cystic clinically. Her past medical history was remarkable for a remote history of treated lymphoma, the details of which were not available. There was no history of apocrine or adenexal tumor elsewhere in the body. Clinically there was no regional lymph node enlargement.

Excisional biopsy of the tumor showed a pink, tan hair bearing fragment of skin measuring 2.7×2.0×1.8 cm. The resection margin was inked. Serial sectioning of the specimen revealed a nodular dermal tumour with focal areas of cystic degeneration filled with a tan coloured paste like material.

Microscopic examination revealed fairly well circumscribed but unencapsulated dermal nodule composed of solid sheets and trabeculae of cells with features of adenocarcinoma (Figure 1A). The cells were large, polygonal with well-defined cell membrane and abundant granular eosinophilic cytoplasm. The nuclei showed moderate pleomorphism with vesicular chromatin and prominent central nucleoli. Abundant mitotic activity and focal necrosis were evident (Figure 1B). Focal involvement of the deep inked resection margin was identified. The overlying epidermis

was unremarkable (Figure 1A). Normal mammary-like glands were not identified in the surrounding tissue. There was no lymphovascular invasion.

The carcinoma cells expressed carcinoembryonic antigen (CEA), Cytokeratin (CK) 7, epithelial membrane antigen (EMA) and Gross cystic disease fluid protein (GCDFP)-15, confirming the apocrine nature of the tumor (Figure 1C-F). The cells were negative for estrogen/progesterone (ER/PR), CK20, leukocyte common antigen (LCA), CK5/6, wilms tumor (WT)-1 and hepatocyte paraffin (HepPar)-1.

Discussion

Primary adenocarcinomas of the vulva are rare compared with vulvar squamous cell carcinomas.¹ Primary vulvar adenocarcinomas are classified into sweat gland carcinomas, primary mammary-like carcinomas of the vulva and extra-mammary Paget's disease. These three tumors share some common histological fea-

Correspondence: Babita Kajal, Department of Pathology and Molecular Medicine, Mc Master University, 1280 Main St W, Hamilton, ON L8S 4L8, Canada.

Tel. +1.905.525.9140.

E-mail: babita.kajal@medportal.ca

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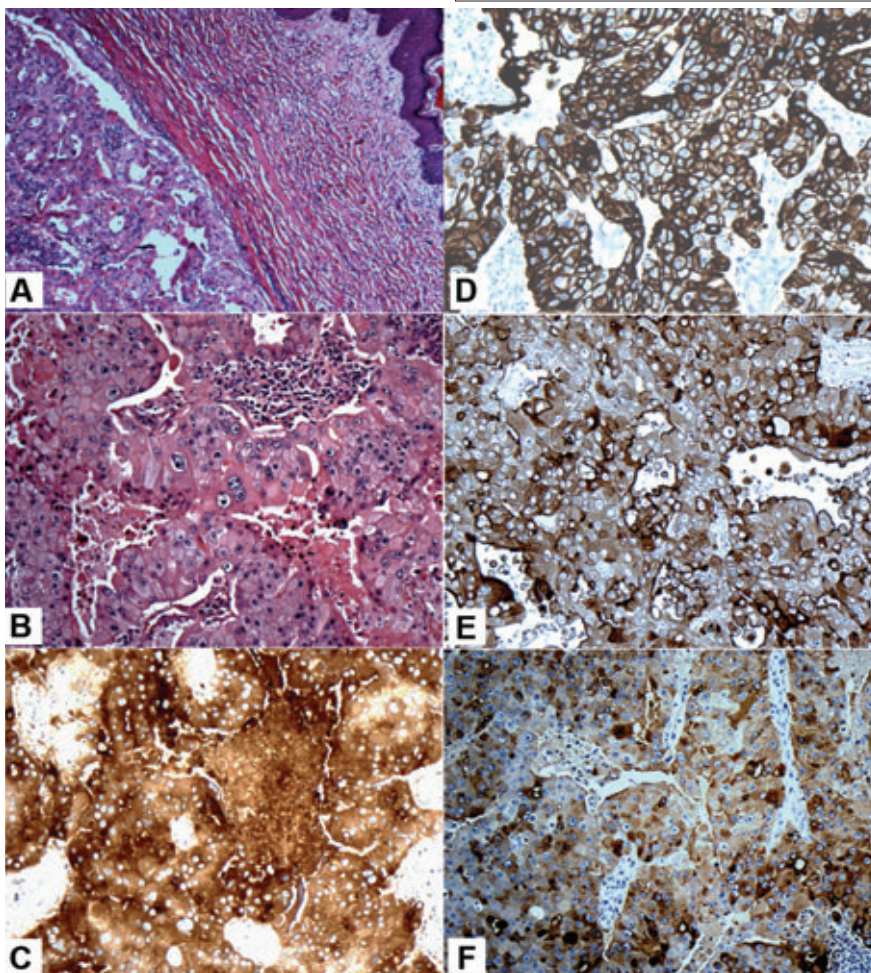


Figure 1. A) Unremarkable epidermis with tumor bulk in dermis 40×; B) Abundant mitotic activity and focal necrosis 100×; C) CEA 100×; D) CK7 100×; E) EMA 100×; F) GCDFP 100×.

tures. Van der Putte and Van Gorp proposed the unifying concept and suggested use of the terms adenocarcinoma of the mammary-like glands of the vulva for these tumors.² Special type of the sweat glands of the vulva, present in highest concentration in the interlabial sulci, when closely studied revealed a specific histology that was different from eccrine, apocrine and mammary glands and at the same time shared features with each of them. It was initially named anogenital sweat gland because it appeared closely related to the eccrine gland,^{3,4} but the term anogenital mammary-like gland was introduced because the microscopic features of mammary glands were obvious.

Rare reported cases of adenocarcinoma with apocrine features have a debatable histogenesis. Origin from the supernumerary breast tissue is unlikely as the milk lines are far away from the anogenital region during embryonic development.⁵ Apocrine carcinoma of the vulva can arise from the native apocrine sweat glands, in which case the tumor is composed of adenopapillary cords and tubules, accompanied by occasional pagetoid components. These tumors are usually negative for ER and PR.^{6,7} Apocrine carcinomas can also originate from the anogenital mammary-like glands in the interlabial sulci, and they usually express ER and PR.^{6,8} The presence of normal mammary-like glands in the vicinity of the tumor and the presence of transition zone between the tumour and the normal mammary-like glands, with positive immunostaining with ER and PR are helpful features to classify vulvar adenocarcinoma as a mammary-like according to the recent literature. In our case, these features were not identified, suggesting origin from native sweat glands rather than anogenital mammary-like glands. Positivity for GCDFP-15, which is localized in the apocrine glands and ducts of normal skin, is essential for pathological diagnosis of apocrine gland carcinoma.⁹

The prognosis of apocrine carcinoma of vulva is poorly understood due to its rarity. Vulvar apocrine adenocarcinoma has a potential for regional lymph node metastasis and metastasis to distant organs has been described.¹⁰⁻¹² Widespread metastases to the heart, lungs, abdominal organs, and bone have been reported.^{8,12,13} The presence of extensive

lymphovascular invasion and metastasis is associated with poor prognosis. In our case, absence of regional lymph node and distant metastasis suggest favorable prognosis. She received surgical interventional only without any hormonal therapy or chemotherapy and she is doing fine as of today with periodical check ups for any potential metastasis. Based upon patient's clinical, physical and radiological examination, there is no breast lesion found, this excludes the possibility of any metastatic disease. There is no specific Immunohistochemistry panel available to differentiate between primary versus metastatic apocrine carcinoma. It all based upon patient's clinical history. There is lot of overlapping of the histological and immunohistochemical features, which makes the distinction very difficult. Before making the definite diagnosis of primary apocrine carcinoma of vulva, through clinical examination and imaging studies are mandatory in order to exclude adenocarcinoma from other primary sites especially (breast, alimentary and female reproductive tract). In our case, patient did not have any suggestion of metastatic disease, so was considered primary apocrine carcinoma of vulva.

Conclusions

In conclusion, our report describes a rare case of primary apocrine adenocarcinoma of the vulva. While we acknowledge the controversial histogenesis, the absence of normal mammary-like glands in the vicinity of the tumor and negative immunostaining for ER and PR are supportive of the native sweat gland origin.

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