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ScienceDirect
Pathology – Research and Practice 205 (2009) 131–135
PATHOLOGY RESEARCH AND PRACTICE
www.elsevier.de/prp

TEACHING CASES

Vulvar apocrine adenocarcinoma: A case with nodal metastasis and intranodal mucinous differentiation

Khaled O. Alsaad, Nidal Obaidat, Valerie Dube, William Chapman, Danny Ghazarian*

Department of Laboratory Medicine and Pathobiology, University of Toronto, and University Health Network, Toronto General Hospital, 200 Elizabeth Street, 11th Floor, Toronto, Ontario, Canada M5G 2M9

Received 27 June 2007; accepted 10 July 2008

Abstract

Primary vulvar adenocarcinomas are rare tumors, and their histogenesis is not fully understood. They are classified into extramammary Paget's disease, sweat gland carcinomas, and "breast-like" adenocarcinomas of the vulva. The latter resemble adenocarcinomas arising in the breast morphologically and immunophenotypically. Rare cases of adenocarcinoma with apocrine features have been reported, and whether these neoplasms originate from the "native apocrine" sweat glands or from "anogenital mammary-like" glands are still debatable. The presence of normal mammary-like glands in the vicinity of the tumor, the transitional malignant morphological features from normal mammary-like glands and the tumor, the breast-like histological features of the tumor, and the expression of estrogen and progesterone receptors generally suggest an origin from anogenital mammary-like glands. Absence of these features points toward native apocrine sweat glands as the source of these neoplasms. In this report, we present a patient who was initially diagnosed with Paget's disease of the right vulva, which was treated by hemi-vulvectomy, and who later presented with primary vulvar apocrine adenocarcinoma with metastasis to the inguinal lymph nodes and intranodal mucinous/colloidal differentiation: a feature, to the best of our knowledge, not reported before. We also reviewed the histogenesis of the vulvar adenocarcinomas, with emphasis on the morphological features that separate the tumors arising from the anogenital mammary-like glands in the vulva from those arising from the native vulvar sweat glands.

Keywords: Vulvar adenocarcinoma; Apocrine; Mucinous; Paget disease

Introduction

Primary vulvar adenocarcinomas are rare tumors. They are classified into sweat gland carcinomas, primary 'breast-like' adenocarcinomas of the vulva, and extra-mammary Paget's disease. Breast-like tissue in the vulva was previously thought to be derived from rudiments of the embryonic milk lines, which extend from the axillae to the groins, a theory that has been recently refuted by van der Putte's studies [18,19]. The breast-like tissue was believed to be the source of benign and malignant breast-like vulvar tumors, such as fibrocystic changes, lactating adenoma, fibroadenoma, phyllodes tumor, hidrocystoma, hidradenoma papilliferum, and *in situ* and invasive adenocarcinoma of mammary-like glands of the vulva, which resemble adenocarcinomas arising in the breast morphologically and immunohistochemically [5,20]. The latter can be of ductal (most common)

*Corresponding author. Tel.: +1 416 340 3839; fax: +1 416 340 5517.
E-mail address: danny.ghazarian@uhn.on.ca (D. Ghazarian).

0344-0338/$ - see front matter © 2008 Elsevier GmbH. All rights reserved.
doi:10.1016/j.prp.2008.07.012

[1,4,10], lobular [13], mixed ductal and lobular [8], or mucinous/colloid [6,23] type. Rare cases of both *in situ* and invasive apocrine adenocarcinoma of vulva have been reported [2,7,9,11,12,14,15,17]. The histogenesis of these neoplasms is still not fully understood, and the question of whether these neoplasms arise from the native apocrine sweat glands or from anogenital mammary-like glands is still debatable. In this paper, we described the features of an unusual case of apocrine adenocarcinoma with metastatic intranodal mucinous/colloid differentiation, and discussed the current understanding of the histogenesis of this tumor.

Case report

An 89-year-old woman presented with irritation for 6 years and a gradually enlarging mass in the left vulva and palpable left inguinal nodes for 1 year. Physical examination revealed an ulcerated fungating mass that involved the left labium majus, clitoris, and the mons. Examination of the breasts and axillae was unremarkable. Her past surgical history includes cutaneous extramammary Paget's disease with dermal invasion of the right vulva, which was treated by right hemivulvectomy and vaginal hysterectomy in 1998, appendectomy for acute appendicitis, and excision of multiple cutaneous basal cell carcinomas. Her medical history was positive for hypertension, rheumatoid arthritis, and osteoporosis. She had no history of breast malignancy, and mammographic findings at the time of presentation were normal. The patient underwent complete vulvectomy and left inguinal lymph node dissection. The surgical specimen contained a firm, tan, nodular tumor that measured 4.7 cm × 4.5 cm. Two lymph nodes that measured 2.2 and 3.5 cm in maximum dimension, respectively, were identified.

Microscopic examination revealed an invasive apocrine adenocarcinoma in the dermis, with a depth of 1.9 cm. The neoplastic cells were of two types: (1) large polygonal cells with abundant, finely granular pale eosinophilic cytoplasm, and indistinct cell membrane (Fig. 1A); and (2) medium size round cells with a small to moderate amount of more eosinophilic cytoplasm and well-defined cell membrane. The tumor cells had large, centrally located, vesicular nuclei, prominent nucleoli, and increased nuclear to cytoplasmic ratio, and were arranged predominantly in solid sheets and nodules and, to a less extent, trabecular and acinar patterns. Occasional multinucleated neoplastic cells were seen. Brisk mitotic activity was present. Variably sized solid nodules of tumor with prominent central necrotic debris mimicking "comedocarcinoma" were present (Fig. 1B). Lymphatic permeation was identified. Normal eccrine and apocrine sweat glands were identified, but there were no normal glands resembling mammary glands in the vicinity of the tumor. Marked stromal desmoplastic reaction was

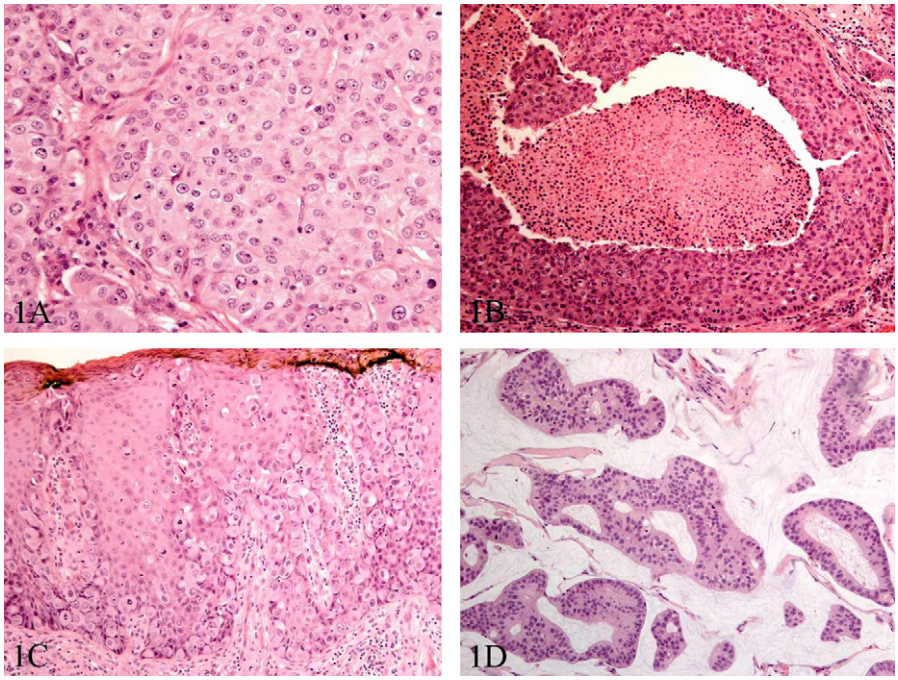

Fig. 1. (A) Tumor cells show apocrine features (hematoxylin and eosin, magnification × 200). (B) Solid nest of tumor with central "comedo"-like coagulative necrosis (hematoxylin and eosin, magnification × 100). (C) Extramammary Paget's disease (hematoxylin and eosin, magnification × 100). (D) Mucinous adenocarcinoma. The tumor cells are arranged in nesting and cribriform pattern floating in pool of abundant mucin (hematoxylin and eosin, magnification × 100).