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Adenocarcinoma with Endometrioid Differentiation of the Labia Minora: A Rare Presentation.

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ABSTRACT

Carcinomas of the vulva are a relatively uncommon gynecologic malignancy of the female genital tract and represent 3–5% of all diagnosed female genital malignancies. Of this primary vulvar glandular neoplasms are still rarer, accounting only 1 % of all malignant neoplasms of the vulva. Endometrioid adenocarcinoma of the labia minora is very rare. We report a 45year old woman presenting with the complaint of whitish discharge admixed with blood per vaginum. On physical examination showed multiple ulcerated nodular lesions with induration in the labia minora largest measuring 3 × 2 cm. An incisional biopsy from the growth revealed adenocarcinoma with endometrioid differentiation extending into the vagina. Among the vulvar cancers, Squamous cell carcinoma is the common histological type. Vulvar adenocarcinomas are rare, arising from adnexal structures, mammary-like glands, vestibular glands and Bartholin's glands. Adenocarcinoma with endometrioid differentiation is extremely rare in which case primary tumour in the uterus and ovaries should be ruled out.

Keywords: Vulva, Labia minora, Vulvar endometrioid adenocarcinoma.

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INTRODUCTION

Cancer of the vulva (also known as vulval or vulvar cancer) most often affects inner edges of the labia majora or the labia minora. Rarely it can arise from clitoris or in the Bartholin's gland. Squamous cell carcinoma comprises 90% of the tumors in vulva [1]. Vulvar adenocarcinomas may originate from adnexal structures, mammary-like glands, vestibular glands, and Bartholin's glands. More common neoplasms include papillary hidradenomas, Bartholin's carcinoma, and Paget's disease of the vulva. Adnexal adenocarcinomas of the vulva are exceedingly rare, accounting for less than 0.1% of all vulvar carcinomas and as a result limited data is available in the literature. The diagnosis of primary vulvar adenocarcinoma remains difficult for pathologists and clinicians [2].

Case Report

A 45-year-old female presented with scanty whitish discharge admixed with blood for the past 3 weeks duration. Physical examination revealed multiple indurated ulcerative nodular lesions on the labia minora largest measuring 3 × 2 cm. On per vaginal examination cervix appeared normal. Growth in the right lip of labia minora extending into lower 1/3rd of vagina and bleeds on touch. There was no history of sexually transmitted diseases, irradiation or tobacco use. Inguinal lymph nodes were not palpable. Ultrasonogram revealed bulky uterus with normal endometrial and myometrial echoes. There was no free fluid in the pouch of Douglas. Both ovaries normal in size and echogenicity. Pap smear study was negative for intraepithelial lesion or malignancy (NILM). Dilatation and curettage was done. Cervix biopsy and endometrial curettings showed chronic cervicitis and proliferative phase endometrium.

Incisional biopsy from the vulvar growth measured 0.5x0.3 cm and bit from lower 1/3rd of vagina measured 0.3x0.2 cm. showed greywhite to greybrown areas. Multiple sections studied revealed infiltration by variable sized irregular shaped glands lined by malignant cells showing stratification and increased mitosis. (Fig. 1). These cells show mild to moderate nuclear pleomorphism with conspicuous nucleoli. (Fig. 2). Diagnosis was adenocarcinoma with endometrioid differentiation of labia minora with infiltration in to the lower 1/3rd of vagina

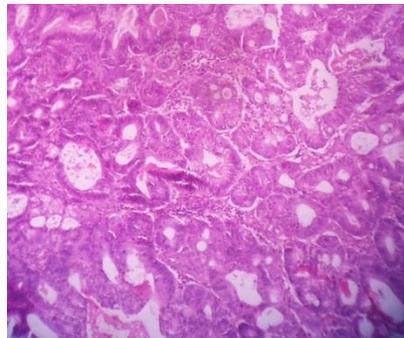


Figure 1: Variable sized irregular shaped glands lined by malignant cells showing stratification and increased mitosis. (hematoxylin and eosin, 10 x)

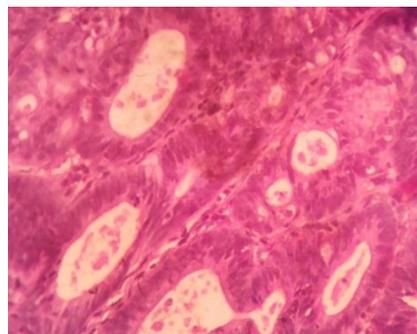


Figure 2: Moderate nuclear pleomorphism with conspicuous nucleoli with endometrioid differentiation (hematoxylin and eosin, 40 x)

DISCUSSION

Most cancers of the vulva are squamous cell carcinomas but tumors arising from labia minora will have diagnostic dilemmas. The rarity and histologic variability may account for the difficulty that will be encountered in the diagnosis of vulval carcinomas. In our case, the possibility of secondary deposits from primary tumors present at other locations, were excluded clinically. Most primary adenocarcinomas of the vulva arise in Bartholin's glands, or are associated with extra-mammary Paget's disease. About 8% of vulvar cancers are adenocarcinomas. Vulvar adenocarcinomas most often start in cells of the Bartholin's glands. These glands are present just inside the opening of the vagina. A Bartholin gland carcinoma is easily mistaken for a cyst (accumulation of fluid in the gland).

So far around 31 cases had been reported in the literature (Stueben BL et al).² Vulvar adenocarcinomas of sweat gland origin have a propensity to spread through the local lymphatics, typically to ipsilateral inguinal lymph nodes [3]. This principle also holds true for apocrine adenocarcinomas arising from the vulva, in which lymphovascular invasion often leads to nodal spread and widespread distant metastasis [4]. With respect to adenocarcinomas of the vulva, several primary and secondary neoplasms must be considered.

Two main primary lesions should be excluded; carcinoma of the Bartholin gland and malignant melanoma [5]. However, this differentiation often makes no difference clinically because the primary treatment modality for both lesions is surgical excision of the vulva with margins of 1 cm or greater [5]. These tumors are resistant to radiotherapy and the benefit of chemotherapy is unknown [6] hence the poor prognosis. Carcinomas originating from Bartholin's glands microscopically may form ducts or have a mucinous (adenocystic) appearance.

However, they are usually readily recognized by the examining surgeon because they arise in the labia minora. By contrast, primary vulvar sweat gland carcinomas arise in the labia majora. It is known that metastasis to the vulva is seen with tumors from the cervix, endometrium, kidney, and the urothelial tract. Since all of these tumors may cause confusion with primary adenocarcinoma of vulva, possibility of secondary deposits from other sites should be excluded as it is more common.

CONCLUSION

In this case, metastatic adenocarcinomatous deposits from the cervix, endometrium and ovary were excluded because of absence of significant findings in pap smear, endometrial biopsy and pelvic ultrasonogram. The histological and clinical findings in this case are consistent with a primary adenocarcinoma with endometrioid differentiation of labia minora which is a rare presentation.

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