

CLOACOGENIC ADENOCARCINOMA OF THE VULVA PRESENTING AS RECURRENT BARTHOLIN'S GLAND INFECTION

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Abstract: Primary adenocarcinoma of the vulva of the cloacogenic type is extremely rare. We report the case of a 49-year-old patient with a 6-year history of recurrent left Bartholin's gland infection and a 1-year history of an ulcerated tumor on the similar lower aspect of the left labium majus. Fine needle biopsy revealed adenocarcinoma and wide local excision with bilateral superficial inguinal lymph node dissection was performed. The patient was well and disease-free at 24 months after the operation. This case provides a good example of why an early biopsy should be performed for any persistent anogenital lesion. Although the reported data on cloacogenic adenocarcinoma are limited, a wide local excision appears to be the most widely used treatment strategy for any vulvar cancer of 2 cm or less in diameter.

Key words: Adenocarcinoma; Vulvar cancer; Bartholin's glands

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Neoplastic diseases of the vulva account for about 4% of all the malignancies of the lower female genital tract. Squamous cell carcinomas comprise approximately 90% of primary vulvar cancers. Adenocarcinomas, which represent about 2–7%, arise mostly from the Bartholin's glands,¹ whereas the cloacogenic histotype was probably first recognized only three decades ago and accounts for a very small percentage. Fewer than 20 cases have been reported. We report a case of vulvar cloacogenic adenocarcinoma presenting initially as a frequently recurrent infection of the left Bartholin's gland.

Case Report

A 49-year-old nulligravid woman had a 1-year history of a painless, solitary, 1.8-cm ulcerated tumor located in the region of the left Bartholin's gland and a 2.3-cm enlarged lymph node in the left groin. Reviewing the past history, left Bartholin's cyst was diagnosed 6 years prior to this visit and the patient was treated with antibiotics. Recurrent infection of *Pseudomonas aeruginosa* had also been noted and she had undergone left marsupialization 2 years prior to this visit. She had a history of *Trichomonas* infection for which she and her spouse were treated. Regular Pap smear tests had been negative for the past 8 years. Fine needle biopsy was performed and

adenocarcinoma was diagnosed. Radical local excision of the tumor was performed with bilateral superficial inguinal lymph node dissection and endometrial curettage. The 2-cm surgical margins and all nodes were negative microscopically. Wound healing was satisfactory without any skin contractions, cosmetic defect or psychosexual dysfunction.

Gross inspection of the resected specimen showed a 1.8 x 1.5-cm polypoid tumor with focal surface ulceration. Microscopic examination showed a well- to moderately-differentiated adenocarcinoma in continuity with the epidermis. Features of adenoma with typical colonic-type mucinous epithelium (Fig. 1) were

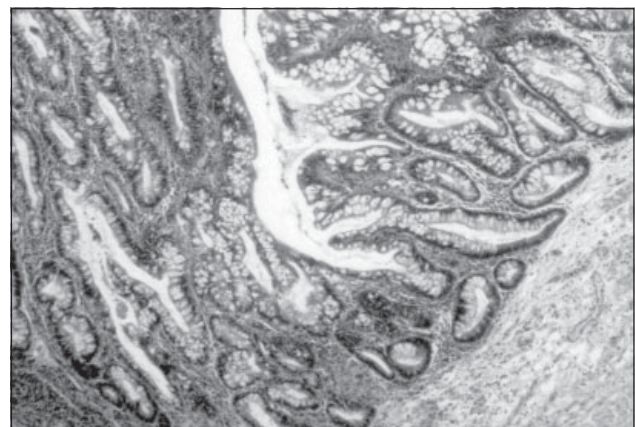


Fig. 1. Transitional area of misplaced cloacal tissue and adenoma (hematoxylin and eosin, x 100).

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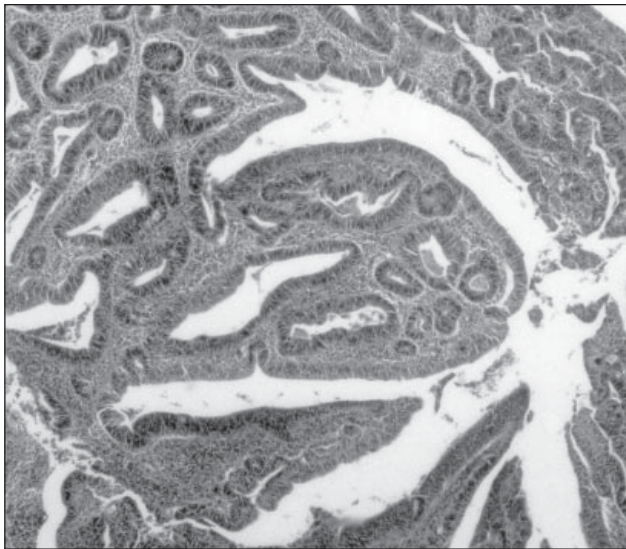


Fig. 2. Transitional area of adenoma and invasive adenocarcinoma (hematoxylin and eosin, x 100).

noted in the tumor margin. Areas of transition to invasive adenocarcinoma comprised of pleomorphic cells arranged in a complex cribriform glandular pattern (Fig. 2) were found. Separated from the tumor were some non-neoplastic normal Bartholin's gland tissues. No areas of apparent transition from normal Bartholin's gland tissues to neoplastic elements were found on histologic study. The tumor cells stained positively with Alcian blue stain and periodic acid-Schiff stain. All lymph nodes and the margins of resection were tumor-free. Endometrial curettage revealed endometrial polyp without atypia or malignancy. The pathological report was consistent with the diagnosis of vulvar cloacogenic adenocarcinoma.

During 2 years of postoperative follow-up, there was no evidence of recurrent disease or other concurrent or subsequent primary neoplasm of similar histological type, and Pap smear and ultrasonographic and sigmoidoscopic studies were negative for tumor. CA-125 levels also remained within the normal range.

Discussion

Cloacogenic adenocarcinoma of the vulva is an extremely rare entity. There is still much controversy regarding its origin and a limited number of case reports.

Initially, Bartholin's gland adenocarcinoma was suspected in this case, as the location of the tumor appeared to be deep within the labium majus and the patient had a history of several preceding episodes of Bartholin's gland infection. Traditionally, treatment of Bartholin's gland cancer requires radical vulvectomy with bilateral inguinal-femoral and pelvic

node dissection.² However, the treatment of vulvar cancer is changing—a less radical, individualized approach has now been adopted for management of this stage I (T1N0M0) presumed Bartholin's gland adenocarcinoma.^{3,4} We used three separate incisions and performed radical local excision together with bilateral superficial groin node dissection. The resection included the tumor per se and extended down to the level of the inferior fascia of the urogenital diaphragm with surgical margins of at least 1 cm. In patients with unilateral lesion, ipsilateral inguino-femoral node dissection is conventional. However, since our patient had a suspicious left groin node of 2.3 cm, bilateral superficial groin nodes above the cribriform fascia were dissected.

The histopathological findings were indicative of cloacogenic adenocarcinoma of the vulva. The treatment strategy for this condition, however, is based on data from a limited number of cases. In patients with no residual disease, postoperative adjuvant inguinal and pelvic radiation is not required.

Reported treatment options include radical vulvectomy and perianal excision⁵; radical vulvectomy with bilateral inguinal lymph node dissection⁶; wide local excision^{6,7}; and modified radical vulvectomy with bilateral inguinal lymph node dissection.⁸ The 2-year disease-free follow-up in this patient suggests that a less aggressive approach (radical local excision with ipsilateral groin node dissection) may be the most suitable, and achieve improved postoperative anatomic and functional status and preserved quality of life in early-stage disease.

In the differential diagnosis of vulvar adenocarcinoma in this patient, the possibility of metastasis was ruled out since the Pap test, sigmoidoscopy and the endometrial curettage were negative for malignancy. The diagnostic criteria for primary Bartholin's gland tumor, as established by Honan in 1897, require that the tumor be in the correct anatomic position, with the location of the tumor deep in the labium majus, with an intact overlying skin and the presence of some normal glandular elements.⁹ Chamlian and Taylor established the diagnostic criteria for a primary Bartholin's gland tumor in 1971 as: (1) areas of apparent transition from normal elements to neoplastic ones on histologic study; (2) the tumor involving the area of Bartholin's gland is histologically compatible with the origin from the Bartholin's gland; and (3) no evidence of primary tumor elsewhere.¹⁰ In this case, there was no evidence of transition from normal Bartholin's gland to adenocarcinoma and hence the above criteria were not fulfilled. On the contrary, a transition from an adenoma with colonic-type mucinous epithelium to invasive adenocarcinoma was clearly identifiable.

Briefly, this tumor showed no direct relationship with the underlying glands, which include the left Bartholin's gland, minor vestibular glands and sebaceous glands. The location of the tumor's origin remained unclear. Based on the similarity of this case to a previous report, the findings of characteristic features of columnar cells with prominent brush border, showing presence of goblet cells and endocrine cells, and demonstrating tubulovillous pattern of enteric adenocarcinoma in this case suggest that cloacogenic adenocarcinoma of the vulva was the correct diagnosis.⁷ The pathogenesis of cloacogenic adenocarcinoma of the vulva is not well understood. The abundance of goblet cells, as in this case, is illustrative of intestinal differentiation in these tumors. The positive tumor stains with Alcian blue stain and periodic acid-Schiff stain found in this case were also noted in previous reports.^{6-8,11,12} However, the speculation that this tumor arises either from cloacal remnants,^{6,8,11} or embryonic nests⁷ is still controversial.

Due to its rarity, primary adenocarcinoma of the vulva of the cloacogenic type has no well-established diagnostic criteria. Most of the reported descriptions include histological features of enteric tissues without evidence of direct contact with the intestine.⁷ Primary vulvar cloacogenic adenocarcinoma, presenting with the characteristics of a Bartholin's gland neoplasm, has not been previously reported in Taiwan.

The current trend in the management of many gynecological malignancies is toward a less radical surgical therapy. Current treatment strategy for vulvar cancer emphasizes a more conservative concept with more concern about morbidity and preservation of quality of life. En bloc radical vulvectomy and bilateral inguinal-femoral lymphadenectomy are now rarely performed. Modern individualized treatment requires careful consideration of the need to reduce complications, minimize psychosexual sequelae and the choice of a less disfiguring surgery without compromising the disease-free survival rate.³ Due to the lack of data available, the principles of management for primary cloacogenic adenocarcinoma of the vulva are similar to those of vulvar cancer and wide local excision is the preferential approach in cases of a superficial lesion.⁶⁻⁸ Our patient had no symptoms of recurrence during regular follow-up visits post-surgery for 2 years. Identifying any sexual dysfunction after the surgery is also a recommended component of appropriate treatment.¹³

In conclusion, relapse of Bartholin's gland infections should be carefully differentiated and treated, especially in peri- and postmenopausal women. Due to the possibility of malignant change, early detection by biopsy is important. As in this case, any suspicious pigmentation, persistent ulceration and long-term pruritus in the vulva warrants early biopsy in order to facilitate differential diagnosis and an appropriate treatment plan.

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