Adenoid Cystic Carcinoma of Bartholin's Gland: Case Report

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Received: 04 Mar. 2015; Accepted: 23 Dec. 2015

Abstract- Primary adenoid cystic carcinoma (ACC) of Bartholin's gland is a rare gynecologic malignancy. We report a case of locally advanced ACC of Bartholin's gland. A 62-year-old presented with left Bartholin's gland carcinoma and underwent left radical vulvectomy, left-sided inguinal-femoral lymph node dissection, posterior pelvic exenteration, and pedicle abdominal muscle flap. On her 3 months follow-up exam she was disease free.Pelvic exenteration for thetreatment of this rare disease in the vulva is a potential curative option.

Acta Med Iran, 2016;54(12):820-822.

Keywords: Adenoid cystic carcinoma; Pelvic exenteration

Introduction

Adenoid cystic carcinoma (ACC) originating from Bartholin's gland is a rare gynecologic malignancy, accounting for <1% of all female genital malignancies and .1-7% of all vulvar carcinomas (1). ACC can occur in many anatomical sites, such as the salivary glands, upper respiratory tract, nasopharynx, breast, uterine cervix and brain (2). The most frequent location of ACC is the minor salivary glands, where this tumor appears to behave as it does in the Bartholin's gland (3).

Although ACC has a tendency to be slow growing, there is a marked predilection for perineural invasion (4). A high rate of recurrence and metastasis to regional lymph nodes, lungs, and bones can also occur. The 10-year survival rate is 59% (4). There are several histopathologic variants of ACC including classic, tubular, or mixed (cribriform, tubular and solid), however,histopathologic pattern has not been found to be predictive for survival (4).

In this report, we discuss a case of locally advanced ACC requiring a posterior exenteration. Including this case, only 4 pelvectomies have been reported in the literature for treatment of this disease (2).

Case Report

A 62-year-old multiparous, postmenopausal woman presented to the outpatient department after her urologist identified a palpable mass on the left side of her vulvar area, associated with intermittent tenderness. The patient did not recall having a mass but did recall a feeling of increased pressure in her vaginal area. Physical examination found a fixed, solid mass about 4 cm x 5 cm over the left posterior labia minor extending laterally, at the site of the Bartholin's gland. The mucosa was intact, and no discharge or bleeding was noted. Pelvic bimanual examination revealed a normal cervix, normal uterine size, and no palpable masses in the adnexa.

The patient's laboratory data on admission, including complete blood counts and kidney and liver functions, were within normal limits. Serum levels of tumor markers CA 125 and CEA were also in the normal range before treatment. The patients MRI showed a left lateral wall enhancing vaginal mass and two intramural fibroids measuring up to 1.7 cm. PET scan showed a metabolically active soft tissue nodule in the left posterior lateral vaginal wall, consistent with a Bartholin's gland carcinoma. There was no evidence of metastatic disease. Pre-operative biopsy was consistent with adenoid cystic carcinoma of the Bartholin's gland.

The patient then underwent a left radical vulvectomy and left sided inguinal-femoral lymph node dissection and the specimen (Figure 1A) with 1cm clearance of the palpable tumor mass, and this was then sent for frozen section analysis of the margins.

The tumor margin was microscopically positive in the rectovaginalseptum, and thus the decision was made to proceed with a posterior exenteration with an end

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colostomy. Due to the size of the pelvic-perineal defect, a pediculized rectus abdominismusculocutaneous flap was created to fill the defect (Figure 1B). Operative time was 8 hours. Total blood loss was estimated at 500cc. The patient's postoperative course was uncomplicated, and she was discharged 8 days postoperatively.

Pathology showed the Bartholin gland tissue to be completely replaced by tumor. This corresponds to an adenocarcinoma forming glandular and cribriform structures with prominent perineural invasion, consistent with vulvar adenoid cystic carcinoma (Figure 1C and 1D). Occasional foci suspicious for lymphovascular space invasion were also seen. Given the location of the tumor and absence of disease in other possible gynecologic sites of origin such as the cervix and ovaries, the findings are compatible with a primary Bartholin gland tumor (3.7 cm diameter). The final surgical margins were free from disease. The left inguinal lymph nodes were negative for metastasis.

She received 5040 cGry in 30 fraction, 5 days a week for 6 weeks due to the extensive perineural involvement. On her 3 months follow-up exam she was disease free.

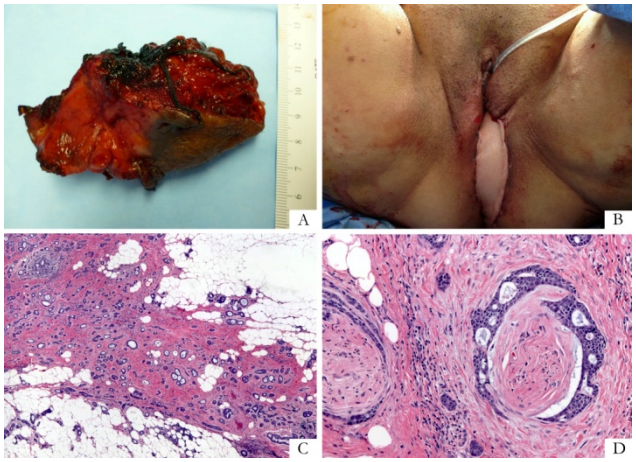

Figure 1. Primary adenoid cystic carcinoma of Bartholin's Glands A: Portion of left vulva and posterior vagina with anarea of retraction indicating the location of the tumor. Suture marked at 12 o'clock. B: After perineal reconstruction with the rectus abdominismusculocutaneous flap. C: adenoid cystic carcinoma infiltrating deep vulvar-vaginal connective and adipose tissue (HE, x40). D: perineural invasion and cribriform structures (HE, x200).

Discussion

ACC is a specific variant of adenocarcinoma of the salivary and mucous glands, which was first recognized in 1859 by Billroth (5). The original diagnostic criteria of ACC of Bartholin's gland was established by Honon in 1897, and in 1972 Chamlain and Taylor re-established the diagnostic criteria to correlate better with findings of advanced tumors (5). Areas of transition from normal to neoplasia must be found on histological findings (5). The tumor must also be compatible with Bartholin's gland's origin, and no evidence of a previous, concurrent, or subsequent primary tumor should be identified (6).

The clinical diagnosis of ACC of the Bartholin's gland includes the tumor located in the Bartholin's gland region, overlying skin intact and tumor located deep in the labia major (7). The clinical symptoms of ACC of Bartholin's gland are nonspecific and easily confused with other diagnoses, such as Bartholin's gland abscess. The symptoms may include a painless lump in the posterior half of the vulva with or without ulceration along with dyspareunia, abnormal bleeding, pruritus and very rarely, vulvar pain (8). The nonspecific symptoms often lead to a delay in both diagnosis and treatment.

Due to the rarity of this tumor, there is no clear consensus regarding the best surgical treatment for ACC of Bartholin's gland. In most cases, either a simple excision or radical vulvectomy with or without lymph node dissection is performed. When comparing recurrence rates for surgery intervention, there is a higher recurrence rate in those that underwent simple excisions (69%) when compared with those that had radical vulvectomies (43%) (8). The initial data from Yang *et al.*, showed that there was a positive surgical margin in 48% of patients who underwent simple excision and 30% in the radical vulvectomy. These results suggest that those individuals that undergo

simple excision are more likely to have positive surgical margins and thus are more likely to have recurrence of thedisease. Shih-Tien *et al*. conducted a review of surgical intervention including all the new cases since 2006. Their data also suggested that radical vulvectomy can reduce local recurrence (46.7%) as compared to simple excision (67.7%) (5). There is no significant difference in the rate of distant metastasis after simple excision (32.3%) and radical vulvectomy (36.7%) (5).

It is unclear whether performing either a unilateral or bilateral inguinal-femoral lymphadenectomy is beneficial since metastatic disease to this area is uncommon. The most common metastatic sites are the bone and lung (6). Reports about the success rates of chemotherapy for ACC are inconclusive. Several chemotherapy agents have been used for this disease with different combinations of Adriamycin, cyclophosphamide, methotrexate, and dactinomycin developed but the response rates to chemotherapy are low, and the duration of response is short (5,10). According to Copeland *et al*., 5-year progress free interval is 47%, and the 5-year survival rate is 71% (7).

Postoperative external beam radiation has been shown to be beneficial in patients with positive resection margins (5). Our patient had negative resection margins;however, her pathology revealed extensive perinueral invasion. ACC has a tendency to recur along nerves and radiation is recommended in head and neck ACC with perineural involvement. The patient in our report received 5040 cGry in 30 fractions, 5 days a week for 6 weeks due to the extensive perineural involvement.

There is no clear consensus as to the proper diagnosis, surgical intervention, and chemotherapeutic treatments to extend patients survival time and reduce the recurrence rate. Reports to date suggest that the single most important factor for cure appears to be surgical resection margins. Thus pelvic exenteration should be considered in patients with ACC of the Bartholin's gland as a possible preferred treatment option in order to surgically clear the disease.

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