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Case report

29-year-old with dyspareunia and vulvar mass: An unusual diagnosis of Bartholin's gland carcinoma

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1. Introduction

Vulvar cancer, including primary Bartholin's gland carcinoma, affects approximately 6190 women yearly with 1200 deaths in 2018 (Siegel et al., 2018). Of these, 0.1% – 7% are Bartholin's gland in origin (Bhalwal et al., 2016; Cardosi et al., 2001). Adenoid cystic carcinoma is an uncommon variant of Bartholin's gland carcinomas, accounting for 10–15% of Bartholin's gland malignancies, with fewer than 90 cases described (Alsan et al., 2011; Bhalwal et al., 2016). Adenoid cystic carcinoma is more commonly a salivary tumor.

Here we present a case of a 29-year-old multigravida with dyspareunia found to have adenoid cystic carcinoma of the Bartholin's gland.

2. Case description

A 29-year-old female presented to her gynecologist's office with six months of gradually worsening dyspareunia. She had noticed two months prior a right-sided “lump” and developed nearly constant discomfort with sitting. She denied additional complaints. She had three uncomplicated vaginal deliveries, no history of sexually transmitted infections and a recent normal pap smear. In the year prior, pelvic ultrasound showed polycystic ovaries, a small leiomyoma, and echogenic myometrium consistent with adenomyosis. Her gynecologist confirmed a mass on examination, obtained a pelvic MRI, and referred her to gynecologic oncology.

Pelvic MRI with IV contrast revealed right-sided asymmetry in the vulvar region, described as a 1.9 cm × 1.5 cm rounded, relatively smooth area. It did not have an aggressive appearance and was hyperintense on T2, atypical for usual pathologies. It was separate from anus and urethra, located lateral to the vaginal wall (Fig. 1).

On examination, she had normal appearing external and internal genitalia, with a firm, 1–2 cm tender mass palpable within the right vulvar Bartholin's gland area and labia majora swelling. The mass was not evident on visual inspection. There was no surrounding nodularity on vaginal or rectovaginal exam.

She underwent an exam under anesthesia with Tru-Cut biopsies of the mass, revealing a pathologic diagnosis of basaloid neoplasm, consistent with adenoid cystic carcinoma. A PET-CT showed no evidence of metastatic disease. She later received a right partial radical vulvectomy and right sentinel lymph node biopsy using Tc-99 injected along the lesion preoperatively. Given the mass was located over 2 cm from midline, only unilateral lymph node dissection was performed. This was followed by a fasciocutaneous pedicled internal pudendal artery perforator flap reconstruction (Fig. 2). The final histopathology was consistent with adenoid cystic carcinoma, described to have an infiltrative growth pattern with predominantly tubular and cribriform component, and focal solid component (< 30%) (Fig. 3). Given disease rarity, pathology was confirmed by an outside gynecologic oncology pathologist. The tumor was 2.4 cm in greatest dimension and without perineural involvement. There was a less than one-millimeter margin at the deep margin from 9 to 10 o'clock and the peripheral margin from 3 to 4 o'clock. Right sentinel lymph node biopsy identified no invasive carcinoma in two inguinal sentinel nodes. During recovery, the medial flap partially dehisced and developed necrosis. She returned to the operating room two weeks after initial operation for debridement and re-excision of surgical margins. No additional carcinoma was identified. Further options offered to the patient included MRI surveillance given difficulty in exams secondary to flap placement versus radiation therapy for prevention of local recurrence. Patient opted for surveillance via yearly MRI monitoring for a minimum of five years.

3. Discussion

Most cases of adenoid cystic carcinoma of the Bartholin's gland present in seemingly benign ways. The most common complaints are burning pain, palpable mass, and swelling (Akbarzadeh-Jahromi et al., 2014; Alsan et al., 2011; Bhalwal et al., 2016; Chamlian and Taylor, 1972; Hsu et al., 2013; Hwang et al., 2012; Nowak et al., 2014).The burning sensation is thought to be secondary to perineural involvement. Initial misdiagnosis or delayed diagnosis may occur in up to 50% of patients (Chamlian and Taylor, 1972) due to the overlap of these complaints with benign conditions, such as cyst or abscess (Chamlian and Taylor, 1972; Hsu et al., 2013) or endometriosis (Akbarzadeh-Jahromi et al., 2014). Adenoid cystic carcinoma has been reported in a large age range (Alsan et al., 2011; Hsu et al., 2013; Yoon, et al., 2015), including the patient presented here who was only 29 years old.

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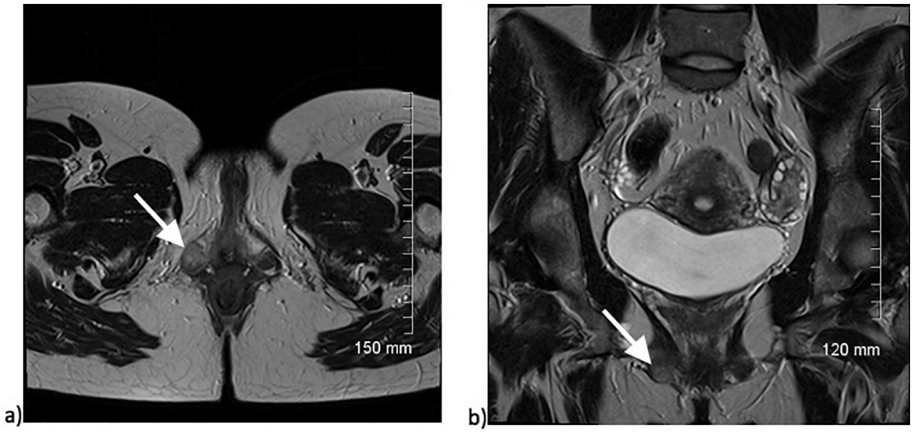

Fig. 1. Axial (1) and coronal (b) T2-weighted images demonstrate mild asymmetric enlargement and subtle hyperintense signal in the right Bartholin's gland (arrows).

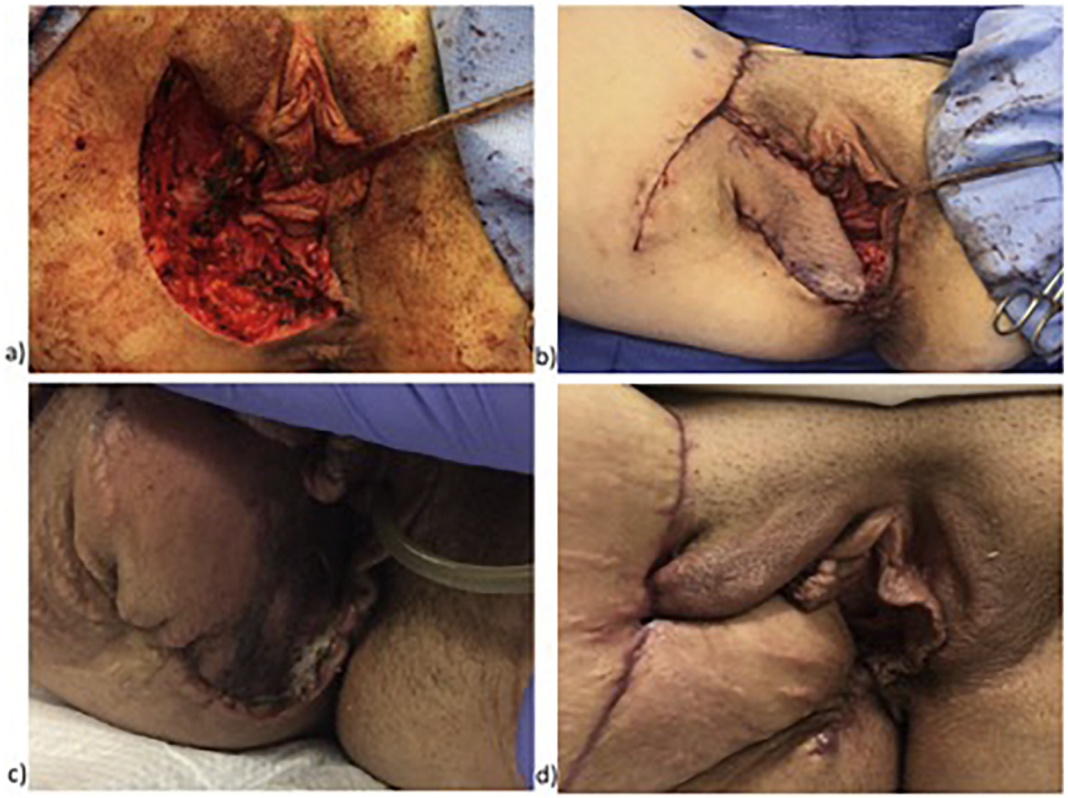

Fig. 2. a) Intraoperative prior to closure from partial radical vulvectomy b) Immediately postoperative from Singapore flap closure c) Medial flap necrosis prior to debridement d) Well healing surgical site from most recent visit.

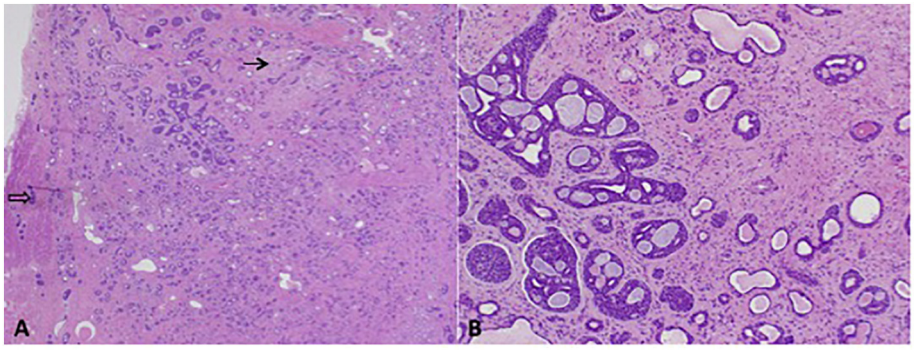

Fig. 3. Adenoid cystic carcinoma. a) Infiltrating neoplasm is adjacent to and dissecting Bartholin's glands (filled arrow) with invasion into skeletal muscle (hollow arrow) (*hematoxylin & eosin, x 20*). c) The neoplasm composed of hyperchromatic basaloid cells with various growth patterns including tubular, cribriform and solid (*hematoxylin & eosin, x 100*).

Classically, Bartholin's gland cancers are diagnosed in older or postmenopausal women. However, adenoid cystic carcinoma of the Bartholin's gland does not show this age predisposition; it is equally distributed beginning mid to late twenties (Alsan et al., 2011). The youngest reported patient was 25 years old. Nearly all patients are parous and cases have been reported during or immediately after pregnancy (Copeland et al., 1986; Murphy et al., 1962). Whether this is a causative factor or secondary to increasing parity with age is unknown.

Diagnosis of Bartholin's gland carcinoma is based on histology and clinical features. Histology must be compatible with Bartholin's gland, with areas of transition from normal to neoplastic elements. The tumor must be located in appropriate anatomic location without evidence of other primary (Chamlian and Taylor, 1972). Adenoid cystic carcinoma of the Bartholin's gland shares classic histologic features with adenoid cystic carcinoma of the salivary gland. It may originate from myoepithelial cells and histology is described as cells in cribriform patterns with mucin filled pseudo-lumens (Di Donato et al., 2017). Most tumors have perineural invasion, which is thought to contribute to its high local recurrence rate. It is unusual in this case to lack perineural invasion, as many consider this a hallmark of the disease. No studies have been performed evaluating preoperative imaging in adenoid cystic carcinoma or general cancers of the Bartholin's gland, although preoperative imaging seems reasonable given its utility in vulvar cancers.

Similarly, therapies for adenoid cystic carcinoma of the Bartholin's gland are based on successes with other vulvar cancer (Di Donato et al., 2017; Yoon et al., 2015). Treatment strategies are based on surgical intervention, simple or radical excision, with or without lymph node dissection. Adjuvant radiation therapy is frequently offered, especially if patient is at high recurrence risk.

While the disease has a 64% survival at 10 years, it tends to be locally aggressive and recurrent (Alsan et al., 2011). Current literature suggests 30% of patients will develop recurrent disease (Alsan et al., 2011), similar to the 40% of patients who develop recurrence after 5 years with non-subtyped Bartholin's gland carcinoma (Bhalwal et al., 2016). Therapy does appear to affect recurrence risk. When considering resection with radical vulvectomy versus simple excision, systematic review by Aslan et al. reports radical vulvectomy patients had more than one recurrence at a rate of 7% compared to 26% with simple excision. Radiation therapy does seem to decrease local recurrence, especially with positive margins. Positive margins had 35% recurrence risk versus 10% with negative margins in the same systematic review.

Treatment for metastatic disease, commonly found in the lungs, bones or liver, varied greatly. On review of case reports, patients received a variety of chemotherapies. One patient's recurrent metastatic disease stabilized after 6 cycles of doxorubicin and cisplatin (Yoon et al., 2015). Use of doxorubicin, cisplatin, cyclophosphamide, dactinomycin, methotrexate, paclitaxel, and docetaxel have all been reported (Alsan et al., 2011; Hsu et al., 2013; Yoon et al., 2015). In a patient who declined chemotherapy described by Hsu et al., tamoxifen was administered after metastases to the lungs with stabilization for four years prior to death from disease. There is not a standard regimen for metastatic or recurrent disease.

There is a lack of data regarding surveillance in adenoid cystic carcinoma of the Bartholin's gland. In this patient, clinical exam alone is ineffective given the reconstruction limits examination of the primary site. For salivary adenoid cystic carcinomas, surveillance is performed via imaging with CT or MRI with baseline scan at 3–6 months after surgery (Hermans, 2008). After discussion with radiology, we elected to use a similar surveillance strategy, with baseline MRI followed by yearly scans.

4. Summary/Conclusion

Given this patient's negative margin status, lack of perineural invasion, negative lymph nodes and lack of metastases, she may be considered optimally surgically resected. While general risk of recurrence in all patients is considered 30%, hers may be lower given lack of risk factors. Even so, she will need regular surveillance, for minimum of 5 years and low threshold for continued surveillance past the 5 year mark given the propensity of this tumor to have long periods of indolence.

This case is important for a number of reasons. There is a possibility of carcinoma even in relatively young patients, even though the risk is rare. Biopsy of such masses, even in this age group, is therefore indicated unless the mass is clearly a Bartholin's cyst or abscess. This is an exceedingly rare cancer and rigorously studied evidence based therapy is not available, but current management including radical resection with lymph node sampling and option for radiation therapy for prevention of local recurrence appears to be a well documented and accepted strategy. Optimal surveillance is not entirely clear, but logic would dictate it is indicated given high recurrence risk.

At last visit six months post operation, patient was recovering well from survey without evidence of disease.

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Conflict of interest statement

None of the authors have conflicts of interest to disclose.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Each author participated in drafting or editing the manuscript. Additionally, Dr. Jackson is the patient's primary oncologist and provided the operative and postoperative photographs. All authors have approved the final article.

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