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A Large Adenoid Cystic Carcinoma of Bartholin Gland Misdiagnosed and Mistreated as a Bartholin Cyst: A Case Report

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ABSTRACT

Introduction Bartholin gland primary cancer occurs scarcely. Adenoid cystic carcinoma of the Bartholin gland is one of the rarest types of primary adenocarcinoma. Treatment delayed can lead to high rate of local recurrence and distant metastasis. The aim of this experimental case study was to discuss a woman with misdiagnosed and mistreated ACC-BG, as a benign situation.

Patient Information In this experimental case study, a 54-year-old woman referred to Joint Committee in Gynecology Oncology Department of Imam Khomeini Hospital of Tehran University of Medical Sciences, Tehran, Iran on Aug 2016 with a large mass within her vaginal distal wall. This woman was undertaken marsupialization for Bartholin cyst. Because of the bizarre manifestation of intervention, the biopsy was performed and revealed as adenoid cystic carcinoma of the Bartholin gland. Therefore, wide local excision and ipsilateral inguinal lymphadenectomy were considered. Adjuvant irradiation was planned.

nificant difference in glucose levels in the first and second fingertip blood drops (p=0.257), while there was a significant difference between glucose levels in the first and second fingertip blood drops with standard venous sample (p<0.05). There was also a significant correlation between 2 drops in expression of glucometry results.

Conclusion Adenoid cystic carcinoma of the Bartholin gland is very rare, but despite its low prevalence, it should be considered especially in the older patient or when the Bartholin cyst is unusually large and sticking to the surrounding tissue to prevent delays in diagnosis and treatment.

Keywords Adenoid cystic carcinoma; Bartholin Gland; Cyst; Adenocarcinoma

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[1] Chemoradiotherapy with irinotecan (CPT-11) for adenoid cystic carcinoma of Bartholin's gland: A case report and review of the literature [2] Liver metastasis in a case of adenoid cystic carcinoma of the Bartholin's gland: A rare presentation [3] Adenoid cystic carcinoma of the Bartholin's gland: Rport of two cases and review of the literature [4] Adenoid cystic carcinoma of the Bartholin's gland: Case report and systematic review of the literature [5] Bartholin's gland carcinoma, malignant melanoma and other rare tumours of the vulva [6] Adenoid cystic carcinoma of Bartholin's gland: A report of five new cases treated with surgery and radiotherapy [7] Adenoid cystic carcinoma of Bartholin's gland: Report of a case and review of literature [8] Analysis of clinical outcomes of patients with adenoid cystic carcinoma of Bartholin glands [9] Adenoid cystic carcinoma of Bartholin's gland clinically mimics endometriosis, a case report [10] Adenoid cystic carcinoma of Bartholin's gland: A case report [11] Adenoid cystic carcinoma of Bartholin's gland-A rare entity likely to be misdiagnosed [12] Adenoid cystic carcinoma of Bartholin gland

Introduction

Adenocarcinoma of the Bartholin Gland (BG) usually is secondary and indeed it is caused by metastasis of other sites of adenocarcinoma. Primary adenocarcinoma of the BG is very scarce approximately 1% to 7% of vulvar cancer [1, 2].

Pure adenocarcinoma, adenosquamous carcinoma, transitional cell adenocarcinoma and adenoid cystic carcinoma are the various type of BG cancer, and the recent subtype, adenoid cystic carcinoma (ACC) comprises about 10% of them [3]. ACC-BG may occur during pre or postmenopausal period of life. It is usually symptomatic and the patients complain of palpable mass accompanying pain or burning sensation [4].

The exact diagnosis is based on pathologic findings. According to this fact that tumors of BG are often secondary and the primary cancers are very rare, particular clinical and pathological criteria must exist for diagnosis primary masses. These criteria for primary BG cancer in general and for ACC as special subtype have been introduced. These criteria are as follows: Tumor anatomical location just in BG depth of major labium, healthy skin cover, the existence of transitional zone from normal glandular structures into tumoral tissue in the pathological examination, and especially for ACC, cribriform pattern of round to oval-shaped cells surrounded with hyaline stroma [5, 6]. Pre-neural involvement is a characteristic pathological finding in ACC, considered the cause of burning sensation [7].

There is still no unit consensus on optimal treatment because of the rarity of this tumor. The cornerstone initial treatment is wide local excision. Although in some cases more radical surgery methods such as hemivulvectomy, total vulvectomy has been used. The therapeutic value of lymphadenectomy is unclear. There is also controversy about adjuvant therapy [8]. Only about 80 cases of ACC-BG have been reported in English language literature so far.

The aim of this experimental case study was to discuss a woman with misdiagnosed and mistreated ACC-BG, as a benign situation.

Patient and Methods

In this experimental case study, a 54-year-old woman referred to Joint Committee in Gynecology Oncology Department of Imam Khomeini Hospital of Tehran University of Medical Sciences, Tehran, Iran on Aug 2016 with a large mass within her vaginal distal wall.

As previous history, she had a small palpable mass on that location about 4 years ago, which suddenly became larger. She had not any sexual intercourse, this and the shame of having to go to the doctor, delayed the diagnosis. Three months before and after mass enlargement, she referred to a non-teaching hospital and marsupialization was performed with the diagnosis of Bartholin gland cyst. Due to the adhesion of the mass to the surrounding tissue and its unusual intra-operational manifestation, her surgeon was done a biopsy, and eventually the histopathologic diagnosis was compatible with adenoid cystic carcinoma of the Bartholin gland (ACC-BG) accompanying surrounding muscular invasion.

Three months later, she came to the department. On her initial physical examination, there was a large painless mass about 6 and 7cm just on the BG location, on the left lateral, extended to the superior one-third of the vaginal wall. It seemed to be very close to the underlying ischium bone and levator ani muscles. Apart from previous marsupialization, the covering mucosa was without lesion, and the covering skin was intact. The ipsilateral and contralateral inguinal lymph nodes were not palpable. In addition, her universal physical examinations were normal. To metastasis evaluation, chest, and abdomen-pelvic computerized tomography were done. Neither in laboratory tests nor in imaging studies, there was evidence of tumoral spreading; the second opinion by expert pathologist endorsed the diagnosis. Hence, Joint Committee planned a radical left hemivulvectomy and ipsilateral lymphadenectomy because of the large size of the mass. Although the rectal mucosa was normal on the examination, the bowel preparedness was considered for possible resection due to being too close to the mass. Examination under anesthesia created the idea that wide local excision with secured surgical margin may be possible despite of its much closed to proximity organs, such as bone, levator ani muscle, and rectum.

Tumoral resection via the mucosal incision was done, there was a very thin layer as a cleavage within dissection, and ultimately, the large tumor had been removed successfully. The mass was about 7 and 8cm in size and penetrated deeply into the fossa iscu-rectal, without adherence to the underlying bone. To get the clear margin, part of Levator Ani muscles also was removed. Because of the large size of the tumor, it was decided to ipsilateral inguinal lymphadenectomy. No enlargement lymph nodes were found but it was completed.

Two small Jackson Pratte devise was placed to drainage blood and discharge for first 24 and 72 hours in vaginal and groin incisions respectively. Her recovery period was uneventful and she was discharged after 5 days of hospitalization. Histopathological examination revealed ACC-BG with tumor size 7cm, the tumor was extremely closed to the surgical margin but not invaded it.

Vaginal margin ulcerated stratified squamous epithelium and congested underlying stroma but

was also free of tumor. All lymph nodes were reactive and free of tumoral invasion. The microscopic characteristics were as follows: Basaloid cells as a glandular structure within the cribriform hyaline stroma. Necrosis was mild but the presence of lymph-vascular space and pre-neural invasion were discovered intensely in almost all cuts of the specimen (Figure 1-A, 1-B).

She received adjuvant irradiation because of the close margin. She was subjected to the follow up every three months. It is now 18 months and there is no evidence in favor of the recurrence.

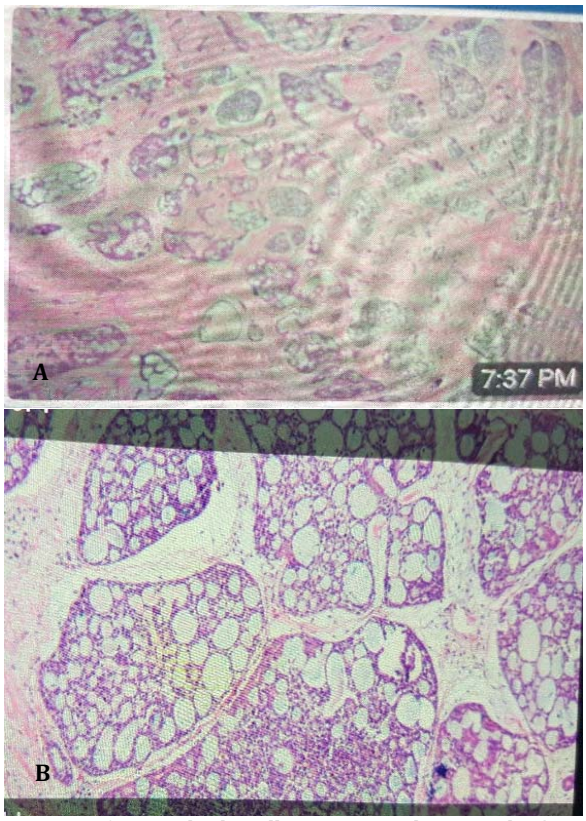

Figure 1) Basaloid cells structured as cribriform glandular pattern within the hyaline stroma, adenoid cystic carcinoma of Bartholin gland, they are pseudo gland because of the myxomatosis manifestation; H&E staining (A, The high magnify picture; B, Myxomatosis manifestation)

Discussion

The aim of this experimental case study was to discuss a woman with misdiagnosed and mistreated ACC-BG, as a benign situation.

ACC is a very rare variant of primary adenocarcinoma which is more common in salivary and breast glands than BG [9]. The most common differential diagnosis is BG-cyst and BG-abscess because in all of them the patient is referred with a palpable mass on the site of BG. Although in abscess formation, the systemic inflammatory response is usually presented accompanying palpable mass and it's suddenly unset distinguishes it.

BG-cyst may also be similar to ACC-BG, particularly, there are no inflammatory symptoms in both, but BG-cyst is completely round and mobile, while ACC-BG penetrates deep into the surrounding tissue which makes it fairly fixed on examination.

In previous reports pre-neural involvement was known as a negative prognostic factor which may lead to higher rate of local recurrences, also the cause of burning sensation in this tumor is nervous invasion [10].

In the present case, despite diffuse pre-neural involvement, she was painless and asymptomatic and the only reason for refers to doctor after four years was the sudden tumoral enlargement. Of course, the threshold of pain and burning in people is different depending on their physical and mental characteristic and these are actually subjective findings. Early detection of asymptomatic mass may be due to alteration in favorite vaginal intercourse but this patient had not any coitus because she was a widow. All of these factors were together led to diagnosis delay. More reports of this situation can provide base data for a large study in the future for introducing optimal treatment option.

ACC-BG has a unique pathologic view, basaloid cells within hyaline stroma, provide a cribriform glandular pattern under microscopic examination [11].

In the present case the special characteristic of the tumor was detected pathologically, and based on her pathologic assertion immunohistochemistry Staining was not necessary to confirm the exact diagnosis.

The rarity of this tumor leads to challenge in selecting optimal treatment and poses the dilemma and controversial points in this issue. There is not any Randomized Controlled Trial (RCT); therefore, there is still no single consensus on best treatment and on adjuvant irradiation as well as chemotherapy [3].

Local recurrence and distant metastasis rate were reported 30% and 31% respectively [12]. Pre-neural invasion and positive surgical margin are negative prognostic factors. Nevertheless, recurrence rate in positive and negative surgical margins have been reported similar in both, so wide local excision seems to be an acceptable option rather than radical surgery. However in positive margin, irradiation is generally recommended to avoid repetitive local recurrences [6].

In the present case despite deeply penetration of the tumor into the Iscue- rectal fossa, and difficult dissection, the cleavage was detected between the tumor and the surrounding tissue, so wide local excision eventually was performed. Ipsilateral lymphadenectomy was considered irrespective of

its unclear therapeutic role. Furthermore, the surgical margin was free of the tumor but it was extremely closed resulted in adopting adjuvant irradiation.

Regardless of the rarity of ACC-BG, it should be considered in each patient with the following features: Older age, long history of mass existence and adhesion or deep penetration into the surrounding tissue. In such these situation, incisional biopsy before surgery is recommended strongly to select optimal treatment option and avoidance of management delay. In the present case, there were all of the risk factors and although biopsy was performed an incomplete marsupialization was also done which can facilitate tumor spreading into the surrounding tissue resulted in difficult dissection on final excision.

The limitations of this research include the lack of doing the frozen section during the first surgery which leads to incomplete removal of the tumor and the suggestion is having a high suspension about malignancy in such situation, for example, a long history of mass existence and adhesion or deep penetration into the surrounding tissue.

Conclusion

Adenoid cystic carcinoma of the Bartholin gland is very rare, but despite its low prevalence, it should be considered especially in the older patient or when the Bartholin cyst is unusually large and sticking to the surrounding tissue to prevent delays in diagnosis and treatment.

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Ethical permissions: The present search followed the ethical standards of Helsinki in 1964 and the consent form for the paper was filled out by the patient before.

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