PAPER DETAILS

TITLE: Adenoid Cystic Carcinoma of Vulva: Case Report

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Olgu Sunumu / Case Report

Adenoid Cystic Carcinoma of Vulva: Case Report

Vulvanın adenoid kistik karsinomu: Olgu sunumu

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Abstract

The adenoid cystic carcinoma (ACC) of the Bartholin's gland (BG) is one of the most uncommon variant of vulvar malignancies representing only 10–15% of cases. Risk factors to the development of ACCBG are still unclear. The symptoms are usually non-specific. In the present study, we present a 61-year-old woman with a vulvar mass evolving since 4 years, without pruritus or pains, without other signs. She initially benefited of an excisional biopsy of the mass returning in favour of a nodular hidradenoma. The evolution after 1 year was marked by the recurrence of the mass. The patient had an ablation of the masse then a total vulvectomy and bilateral lymph node dissection because pathology showed ACC of Bartholin's gland with positif surgical margins. She received adjuvant external beam radiation to the pelvis and on the operating bed. Patient's evolution was marked by complete remission maintained after 4years follow-up. Adenoid cystic carcinoma of vulva is an extremely rare, slowly progressing neoplasm mostly involving the Bartholin's gland. The usual treatment includes wide excision and adjuvant radiotherapy (if required). There may be late local and distant recurrence.

Keywords: Adenoid cystic carcinoma, Bartholin's gland, Surgery, Radiotherapy, Chemotherapy, Surgical margins

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Bartholin'in bezinin (BG) adenoid kistik karsinomu (ACC), vakaların sadece% 10-15'ini temsil eden en sık görülen vulvar malignitelerden biridir. ACCBG'nin gelişiminde risk faktörleri hala belirsizdir. Belirtiler genellikle spesifik değildir. Bu çalışmada biz 4 yıldan beri vulvar kitlesi olan kaşıntı, ağrı ve diğer hiçbir belirtisi olmayan 61 yaşındaki kadın hastayı sunuyoruz. Başlangıçta nodüler hidradenom lehine dönen kitlenin eksizyonel bir biyopsisinden faydalandı.

1 yıl sonraki değerlendirme kitlelerin tekrarı ile işaretlendi. Hastada total bir vulvektomi ve iki taraflı lenf nodülü diseksiyonu ile ablasyon yapıldı, çünkü patoloji, Bartholin'in bezinde positif cerrahi sınırlara sahip ACC olduğunu gösterdi. Pelvise ve ameliyat yatağına adjuvan dış ışın radyasyonu verildi. Hastanın değerlendirilmesi, 4 yıllık takipten sonra devam eden tam remisyon ile işaretlendi. Vulva adenoid kistik karsinomu, çoğunlukla Bartholin bezini içeren son derece nadir, yavaş ilerleyen bir neoplazmdır. Genel tedavi, geniş eksizyon ve adjuvan radyoterapiyi içerir (gerekirse). Geç yerel ve uzak nüks olabilir.

Anahtar Kelimeler: Adenoid kistik karsinom, Bartholin bezi, Cerrahi, Radyoterapi, Kemoterapi, Cerrahi sınırlar

INTRODUCTION

Adenoid cystic carcinoma of vulva (ACC-vulva) is an extremely rare entity with fewer than 100 cases reported in the literature so far (1). In general, ACC affects the exocrine glands. First described by Theodor Billroth (1856), the currently accepted terminology ACC was proposed by Foote and Frazell (1953) (2). Fifty-eight percent of these tumours occur in the oral cavity, major and minor salivary glands, palate, floor of the mouth, gums, lips, tongue and pharynx. Typically, it consists of small basaloid cells with a solid cribriform pattern or epithelial cells with a tubular growth pattern in histology (3). The occurrence of ACC is fairly scarce in the vagina, and , there is limited literature on ACC-vulva. The present report intends to share 1 case of ACC that occurred in the vagina in a 61 year old woman.

CASE PRESENTATION

A Sixty-one-year-old woman with menopause, presented with a swelling on the right posterior labia minor associated with intermittent tenderness. The patient recalled that the

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mass had slowly increased in size from 2cm over a period of 4 years. Physical examination found an indurative, fixed, irregular margin mass of about 5 cm over the right posterior labia minor, just at the site of Bartholin's gland. The mucosa was intact and no pus discharge or bleeding was found. Pelvic bimanual examination revealed a normal cervix, normal uterine size, and no palpable masses in the adnexa. An excisional biopsy by local hospital revealed a nodular hydradenoma.

The evolution after 1 year was marked by the recurrence of the mass. A simple excision was arranged first, The pathology examination showed an adenoid cystic carcinoma predominantly cribriform infiltrating focal soft tissues with perineural neoplastic invasion and without vascular emboli arising from Bartholin's gland (figure.1,2).

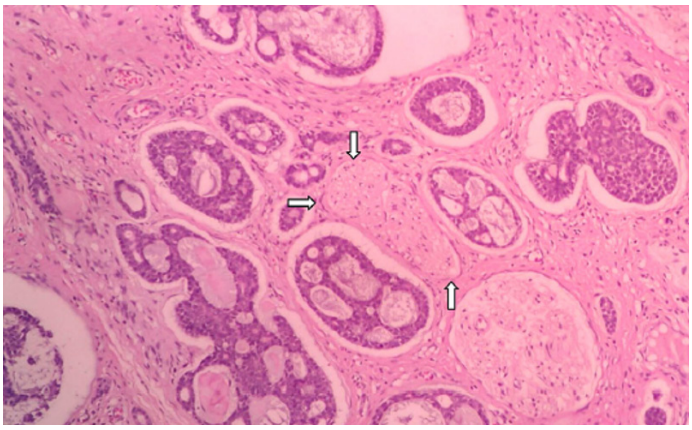

Figure 1. Histopathological picture showing neoplasm with cribriform pattern. Tumour cell nests are seen surrounding a nerve fibre (marked by arrows), indicating perineural infiltration

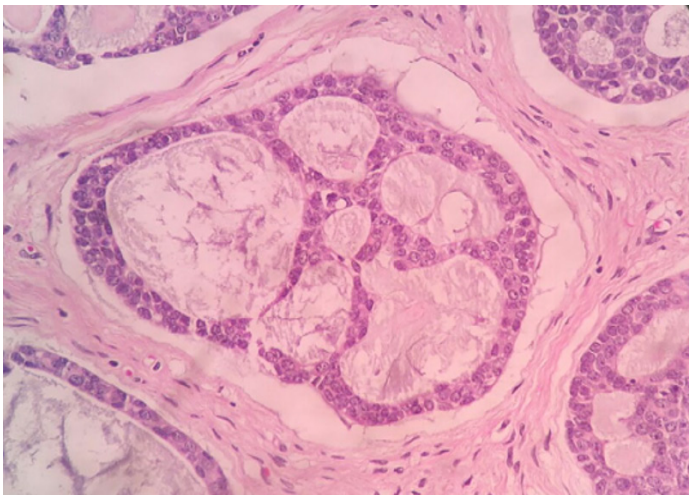

Figure 2. High power view (940) of cribriform patter

Surgical resection limits were at 1 mm of carcinoma. The patient was then referred to our institution and the decision at the multidisciplinary consultation meeting was a surgical revision.

extension assessment was negative so the patient underwent total vulvectomy with bilateral inguinal dissection with resection of a pararectal mass of 3 cm, the histological aspect of the vulvectomy patch was in favour of 2 mm residual of adenoid cystic carcinoma located distant from the resection limits, the pararectal mass: seat of 2.5 * 1.5 cm of adenoid cystic carcinoma infiltrating the pararectal soft tissues with perineural neoplastic invasion and arriving at the confines of surgical resection, right and left curage were negative.

Postoperative pelvic MRI revealed an operative remodeling of the vulvo-anal region, with bilateral inguinal lymphocele. An adjuvant external radiotherapy, at the total dose of 66Gy, was delivered to the patient in 2 series: 1st on the pelvis and a boost on the operating bed (Figure. 3).

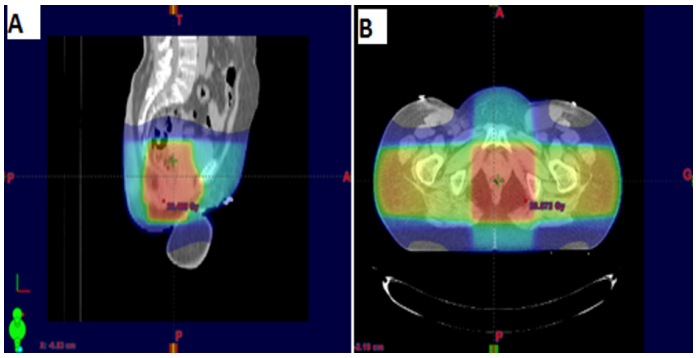

Figure 3. 3D conformational radiotherapy of the adenoide cystic carcinoma of vulv showing the dose's distribution of the Boost (A: Sagittal CT scan image; B: Axial CT scan image)

The patient has been in complete remission for 4 years with adverse effects suche as a post-radiation proctitis and lymphoedema of the two lower limbs.

DISCUSSION

Adenoid cystic carcinoma of vulva usually originates from the Bartholin's gland, but may rarely occur in the rest of the vulva. ACC constitutes only 10 % of Bartholin's gland carcinomas, which in turn, constitute only 0.1–7 % of vulval malignancies (4).

Theodor Billroth was the first to describe ACC as cylindromas in his histological studies in 1856. The incidence of ACC is not very high, in general (5). These are usually observed in the salivary gland. The average age of onset was 57.4 years old, and approximately 60% of patients are women (6). Typical symptoms are similar to those of a Bartholin's gland abscess, including a painless lump in the posterior half of the vulva with or without ulcerations and dyspareunia, abnormal bleeding, pruritus, and rarely, vulvar pain (7). Half of the cases of carcinoma of Bartholin's gland were initially clinically misdiagnosed as cysts or abscesses (8). The clinical diagnosis of a Bartholin's gland tumor includes the tumor located in the Bartholin's gland region, overlying skin intact, tumor located deep in the labia major, normal glandular elements present on histology, areas of apparent transition from normal to neoplastic elements, histological tumor type consistent with the Bartholin's gland origin and no evidence of a previous, concurrent,

or subsequent primary tumor of similar histologic type elsewhere (9). When the tumor progresses, the overlying skin may become ulcerated (10).

The preoperative diagnosis is very hard for clinicians, in general, and most cases are dependent on biopsy. The treatment aspect of this tumor was chiefly surgery or/ and coupled with radiotherapy. There is currently no consensus regarding the optimal surgical treatment for ACC of Bartholin's gland. Both simple excision and radical vulvectomy with or without lymph node dissection have been performed. According to the experience of Lelle et al, if an adequate surgical margin can be achieved, a more conservative surgical procedure with adjuvant radiationmay be reasonable (11). According to the review by Yang et al, 68.9% of patients who had a simple excision had recurrences compared with 42.9% of patients who had a radical vulvectomy. Although the information on the status of margins at the initial surgery was incomplete, the positivity of resection margin was 48% in the simple excision group and 30% in the radical vulvectomy group (12). Correspondently the cas of the patient of this presentation. Radical vulvectomy can reduce local recurrence, but it has no impact on distant metastasis.

Guidelines for postoperative chemotherapy and chemoradiotherapy are not established, despite the frequency of microscopically positive surgical resection margin relatively high. Adjuvant radiation therapy seems to lower the incidence of local recurrence in patients with positive resection margins. Rosenberg et al. (13) and Copeland et al. (14) reported the benefits of postoperative external beam radiation for patients with positive margins. Which is the case of our patient with positive margin who had received an adjuvant external beam radiation. Other adverse prognostic factor for local recurrence reported by Alsan et al. (15) is the presence of neural invasion.

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Conflict of Interest: *The authors declare that they have no competing interest.*

CONCLUSION

Adenoid cystic carcinoma is a rare tumor, usually part of salivary tumors of which it represents 10%. This kind of cancer is characterized by the absence of pain, destruction local structures, and a high rate of recidivism local and metastatic spread.

There is currently no consensus regarding the optimal surgical treatment. Guidelines for postoperative chemotherapy or chemoradiotherapy are not established. Adjuvant radiation therapy seems to lower the incidence of local recurrence in patients with positive resection margins, based on retrospective studies and case reports. Chemotherapy as adjuvant treatment is still under evaluation.

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