Case Reports in Oncology

Case Rep Oncol 2024;17:1282–1288
DOI: 10.1159/000541967
Received: August 8, 2024
Accepted: October 9, 2024
Published online: November 12, 2024

Case Report

Basosquamous Carcinoma Arising from the Vulva: A Case Report

Fatma Saadallah[a, b] Ayoub Ghazouani[a, b] Ines Zemni[a, b] Haykel Turki[a, b] Houyem Mansouri[a, c] Ghada Sahraoui[a, d] Tarek Ben Dhiab[a, b]

[a]Faculty of Medicine of Tunis, University of Tunis Manar, Tunis, Tunisia; [b]Department of Surgical Oncology, Salah Azaiez Institute, Tunis, Tunisia; [c]Department of Surgical Oncology, Jendouba Regional Hospital, Jendouba, Tunisia; [d]Department of Pathology, Salah Azaiez Institute, Tunis, Tunisia

Keywords

Basosquamous carcinoma · Metatypical basal cell carcinoma · Vulvar cancer · Surgery · Case report

Abstract

Introduction: Basosquamous carcinoma (BSC) of the vulva is an uncommon tumor that primarily consists of basal cell carcinoma with squamous differentiation. Also known as metatypical basal cell carcinoma, BSC is typically classified as a skin cancer and represents only 2% of non-melanoma skin malignancies. This type of carcinoma has a poorer prognosis than basal cell carcinoma due to its increased local aggressiveness and metastatic potential. ***Case Presentation:*** We present the case of a 59-year-old woman with a 3-year history of a slow-growing and painful vulvar lesion. Clinical examination revealed a 35-mm nodular, ulcerating, and non-pigmented lesion located on the labia majora. A biopsy confirmed the diagnosis of BSC, and staging assessments indicated no evidence of metastasis. The patient underwent partial radical vulvectomy and sentinel lymph node biopsy, with histological analysis revealing distinct features characteristic of BSC, including basaloid cell islands and areas of significant squamous differentiation. The excision margins were tumor-free, and all six lymph nodes examined were negative for metastases. Regular surveillance for 6 months was conducted without signs of recurrence. ***Conclusion:*** After reviewing the literature, this case represents the sixth documented instance of vulvar BSC. Compared to basal cell carcinoma, BSC has a poorer prognosis, with a higher potential for recurrences and metastases. While basal cell carcinoma is much more prevalent among vulvar and skin malignancies, recognizing the squamous differentiation component is crucial for ensuring wider margins during surgical excision.

Correspondence to:
Fatma Saadallah, fatma.saadallah@fmt.utm.tn

Case Rep Oncol 2024;17:1282–1288
DOI: 10.1159/000541967

Introduction

Basal cell carcinoma (BCC) is a common skin cancer; however, it accounts for less than 1% of all BCC cases in the vulva [1]. Basosquamous cell carcinoma (BSC) is a metatypical form of BCC characterized by a combination of histologic features from both BCC and squamous cell carcinoma (SCC). BSC is rare in the vulva [2]. The etiology of BCC commonly includes factors related to ultraviolet exposure, while hypotheses for vulvar localization have implicated syphilis, chronic irritation or infection, trauma, arsenicals, and radiotherapy [3]. Clinical examination alone cannot differentiate BSC from BCC; definitive diagnosis relies on histology often aided by immunohistochemistry. We report the case of a woman presenting with BSC of the vulva at the age of 59. To the best of our knowledge, this is the sixth reported case.

Case Presentation

A 59-year-old woman, gravida 3 para 2, with a history of hypertension, type 2 diabetes, dyslipidemia, and hyperuricemia, was referred to our cancer institute for a 3-year history of a slow-growing and painful vulvar lesion.

Physical examination revealed a 35 mm nodular, ulcerating, and non-pigmented lesion on the anterior part of the right edge of the labia majora that did not extend to the urethral meatus. No lichen lesions were observed. The cervico-vaginal examination showed no abnormalities, and there were no palpable inguinal lymph nodes. A biopsy of the vulvar lesion was performed, confirming BSC. A cervical smear was also conducted and showed no abnormalities. Staging assessments revealed no evidence of metastasis.

The patient underwent a partial radical vulvectomy with sentinel lymph node biopsy from the right groin. The macroscopic examination revealed a 3-cm nodular, tender, and non-pigmented lesion arising from the labia majora (Fig. 1). Microscopically, the epidermal carcinomatous proliferation organized into well-defined lobules that ulcerated the surface and infiltrated the reticular dermis. The tumor cells were basaloid with nuclei exhibiting a palisading arrangement at the periphery of the lobules. Mitoses were rare, and no lymphovascular invasion was identified. In some areas, squamous differentiation was observed within these lobules, with tumor cells forming clusters surrounded by layers of keratin. The histological examination revealed distinct features characteristic of BSC, including prominent basaloid cell islands and areas demonstrating significant squamous differentiation. These features were sufficiently evident to support a definitive diagnosis without immunohistochemical analysis (Fig. 2).

The excision margins were tumor-free. Six lymph nodes were identified in the lymph node biopsy sample; all were negative for metastases. The Responsible Clinical Panel (RCP) has concluded that no additional adjuvant therapy is necessary. The patient has been undergoing regular surveillance for 6 months with no signs of recurrence.

Discussion

Vulvar cancer represents only 3%–5% of female gynecological cancers and is predominantly of squamous cell type (90%–92% of cases) [4]. BSC is a non-melanoma skin neoplasm first described in 1894 by Beadles [5–7]. The majority of BSC cases (80%) are located in the head and neck region, with only 5 cases involving the vulva documented in literature: 3 cases described by Stewart et al. in 1960, 1 case reported by Kimball et al. in 2006, and another by Hughes et al. in 2021 [8–10].

Case Rep Oncol 2024;17:1282–1288
DOI: 10.1159/000541967

Fig. 1. Partial radical vulvectomy showing a 3-cm white and yellow tumor arising from the labia majora.

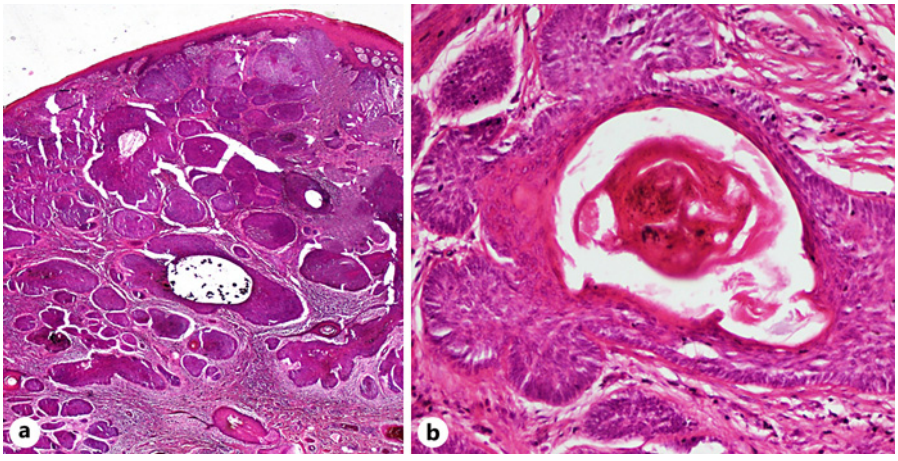

Fig. 2. BSC of the vulva: microscopic examination (hematoxylin-eosin). **a** Grossment ×40: carcinomatous proliferation exhibiting both basaloid and squamous differentiation arranged in nests and nodules. **b** Nests of basaloid cells with peripheral nuclear palisade, centered by foci and squamous cells with keratin lamellae.

Clinically, BSC does not present any particularities; diagnosis is commonly made through biopsy [11]. It often manifests as an indolent ulcerated lesion with or without loco-regional lymphadenopathy.

Histology plays a crucial role in confirming the diagnosis since this tumor type contains both basal cell and squamous cell components with a transition zone between them. Immunohistochemical studies are positive for Ber-EP4, AE1, and AE3 markers in the basal cell area and AE1, AE3, and CAM5.2 markers in the squamous area. The transition zone

Case Rep Oncol 2024;17:1282–1288
DOI: 10.1159/000541967

Fig. 3. Timeline summarizing the main events of the case report.

predominantly shows reduced marking of Ber-EP4 correlating with the transition from basal to squamous zones aiding differentiation from other diagnoses such as collision tumors [12, 13].

Case Rep Oncol 2024;17:1282–1288
DOI: 10.1159/000541967

The prognosis for BSC is more serious compared to BCC due to its metastatic potential; recent studies indicate that, while BSC recurs less frequently than skin SCCs overall, its metastatic potential remains significant. Bowman et al. reported a prevalence of metastatic disease at the presentation of 7.4% for BSC compared to less than 0.1% for BCC and 0.87% for SCC; additionally, BSC exhibits local aggressiveness comparable to pure SCC and BCC [14–16].

In vulvar carcinoma specifically, various studies investigate multiple biological markers as prognostic factors; markers such as p63, Ber-EP4, and EMA can provide insights into biological behavior and treatment response in vulvar cancers specifically [5, 17, 18]. Because of its rarity, there is no consensus on managing both skin and vulvar BSC; however, surgery remains first-line treatment. For skin BSC, authors recommend wider excision margins than those typically used for BCC to achieve better disease control [5, 16]. Sentinel lymph node biopsy is recommended by certain authors; however, there are insufficient data in the literature to assess its benefits [5]. Radiotherapy can be indicated if surgical treatment is not feasible initially or in case of positive surgical margins without the possibility of re-resection [5].

Chemotherapy is restricted to locally advanced forms; immunotherapy may be discussed within multidisciplinary teams for patients who have exhausted other options [5]. The patient's clinical presentation and the predominant histological features, BCC or SCC, are important factors in this decision. Additionally, the use of immune checkpoint inhibitors has gained traction in treating various skin cancers, including BSC, especially for patients who have exhausted other treatment options [19].

Conclusion

BSC represents a rare entity within skin cancers in the vulvar region; this case report highlights the importance of accurate histological diagnosis essential for differentiating BSC from other malignancies such as BCC and SCC. As BSC exhibits more aggressive behavior compared to typical BCC, understanding its prognosis and treatment options is vital. Current treatment strategies primarily involve surgical excision with considerations for wider margins in certain cases while emerging therapies such as immunotherapy may provide additional options for advanced or recurrent disease. Future research should focus on establishing standardized management protocols while exploring novel therapeutic agents' efficacy in improving patient outcomes; given its rarity collaboration among multidisciplinary teams remains crucial for optimizing care tailored to patients' specific needs. This case adds to the limited literature on vulvar BSC emphasizing the need for continued investigation into its biological behavior therapeutic strategies.

A timeline summarizing key events related to this patient's diagnosis and treatment has been included as Figure 3 for enhanced readability.

Statement of Ethics

This case study was reviewed and approved by the Local Ethic Committee of Salah Azaiez Institute, Tunis, Tunisia, on May 22, 2024, Approval No. ISA/2024/13. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000541967).

Conflict of Interest Statement

The authors have no conflict of interest to report.

Funding Sources

The authors received no financial support for the research, authorship, and/or publication of this article.

Author Contributions

Turki Haykal and Ghazouani Ayoub contributed to the collection of patient data. Saadallah Fatma, Zemni Ines, Mansouri Houyem, and Ben Diab Tarek collaborated on the literature search related to the manuscript and the writing of the manuscript. Sahraoui Ghada was responsible for photographing surgical specimens, capturing images with the microscope, and describing the various figures.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

References

1 Namuduri RP, Lim TY, Yam PK, Gatsinga R, Lim-Tan SK, Chew SH, et al. Vulvar basal cell carcinoma: clinical features and treatment outcomes from a tertiary care centre. Singapore Med J. 2019;60(9):479–82. https://doi.org/10.11622/smedj.2019014
2 Mercuț R, Georgescu C, Popescu SM, Ciurea M, Scrieciu M, Georgescu CC. Face basosquamous carcinoma, a case report. Curr Health Sci J. 2014;40(4):277–80. https://doi.org/10.12865/CHSJ.40.04.09
3 Asuman C, Ozlem A, Burçak T, Onder P. An unusual location of basal cell carcinoma: the clitoris and the vulva. Indian J Dermatol. 2008;53(4):192–4. https://doi.org/10.4103/0019-5154.44795
4 Novak E, Berek JS. Novak's gynecology. In: Holschneider CH, Berek JS, editors. Philadelphia: Lippincott, Williams and Wilkins; 2002. p. 303–10 Vulvar cancer. 13th.
5 Murgia G, Denaro N, Boggio F, Nazzaro G, Benzecry V, Bortoluzzi P, et al. Basosquamous carcinoma: comprehensive clinical and histopathological aspects, novel imaging tools, and therapeutic approaches. Cells. 2023;12(23):2737. https://doi.org/10.3390/cells12232737
6 Shukla S, Khachemoune A. Reappraising basosquamous carcinoma: a summary of histologic features, diagnosis, and treatment. Arch Dermatol Res. 2020;312(9):605–9. https://doi.org/10.1007/s00403-020-02058-1
7 Tan CZ, Rieger KE, Basosquamous Carcinoma SKY, Directions F. Controversy, Advances. Dermatol Surg. 2017;43(1):23–31. https://doi.org/10.1097/DSS.0000000000000815
8 Marcus SL. Basal cell and basal-squamous cell carcinomas of the vulva. Am J Obstet Gynecol. 1960;79(3):461–9. https://doi.org/10.1016/0002-9378(60)90244-1
9 Kimball KJ, Straughn JM, Conner MG, Kirby TO. Recurrent basosquamous cell carcinoma of the vulva. Gynecol Oncol. 2006;102(2):400–2. https://doi.org/10.1016/j.ygyno.2006.02.039
10 Hughes NJ, Ntalianis KM, Razvi K, Morgan M, Marzouk S, Sundaresan M, et al. A case of vulval basosquamous basal cell carcinoma with possible pelvic lymph node metastases. Eur J Gynaecol Oncol. 2021;42(4):817–23. https://doi.org/10.31083/j.ejgo4204124
11 Garcia C, Poletti E, Crowson AN. Basosquamous carcinoma. J Am Acad Dermatol. 2009;60(1):137–43. https://doi.org/10.1016/j.jaad.2008.09.036
12 Fotiadou C, Apalla Z, Lazaridou E. Basosquamous carcinoma: a commentary. Cancers. 2021;13(23):6146. https://doi.org/10.3390/cancers13236146
13 Volkenstein S, Wohlschlaeger J, Liebau J, Arens A, Lehnerdt G, Jahnke K, et al. Basosquamous carcinoma-A rare but aggressive skin malignancy. J Plast Reconstr Aesthet Surg. 2010;63(3):e304–6. https://doi.org/10.1016/j.bjps.2009.05.058

Case Rep Oncol 2024;17:1282–1288
DOI: 10.1159/000541967

14 Bowman PH, Ratz JL, Knoepp TG, Barnes CJ, Finley EM. Basosquamous carcinoma. Dermatol Surg. 2003;29(8):830–3. https://doi.org/10.1046/j.1524-4725.2003.29217.x
15 Oldbury JW, Wain RAJ, Abas S, Dobson CM, Iyer SS. Basosquamous carcinoma: a single centre clinicopathological evaluation and proposal of an evidence-based protocol. J Skin Cancer. 2018;2018:6061395. https://doi.org/10.1155/2018/6061395
16 Ciążyńska M, Sławińska M, Kamińska-Winciorek G, Lange D, Lewandowski B, Reich A, et al. Clinical and epidemiological analysis of basosquamous carcinoma: results of the multicenter study. Sci Rep. 2020;10(1):18475. https://doi.org/10.1038/s41598-020-72732-x
17 Garganese G, Inzani F, Fragomeni SM, Mantovani G, Della Corte L, Piermattei A, et al. The vulvar immunohistochemical Panel (VIP) project: molecular profiles of vulvar squamous cell carcinoma. Cancers. 2021;13(24):6373. https://doi.org/10.3390/cancers13246373
18 Sahin TK, Rizzo A, Aksoy S, Guven DC. Prognostic significance of the royal marsden hospital (RMH) score in patients with cancer: a systematic review and meta-analysis. Cancers. 2024;11(10):16. https://doi.org/10.3390/cancers16101835
19 Zelin E, Mazzoletti V, Cavallo F, Nardello C, Corio A, Toffoli L, et al. Treatment of locally advanced and metastatic basosquamous carcinoma, navigating among sonic hedgehog pathway inhibitors, immune checkpoint inhibitors, chemotherapy, and radiotherapy: a case series and literature review. Australas J Dermatol. 2024;65(2):103–13. https://doi.org/10.1111/ajd.14182