BASAL CELL CARCINOMA IN AN UNUSUAL SITE - VULVA

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ABSTRACT

Basal cell carcinoma (BCC) is the most common malignancy of the skin, but BCC of the vulva is a rare entity. BCC of vulva accounts for less than 5% of all vulvar neoplasms and less than 1% of all basal cell carcinomas. Vulvar basal cell carcinoma has a low propensity for metastatic spread but has a high chance of local recurrence after simple excision. We report a 50-year-old woman presenting with pruritus vulva. Physical examination revealed a 2.5 x 1.5 cm hyperpigmented plaque on the left labium majus. The histopathology was consistent with superficially invasive basal cell carcinoma. The patient underwent radical local excision including the clitoris and remained disease free at postsurgical follow-up after eight months.

KEYWORDS
Basal Cell Carcinoma, Radical Local Excision, Vulva.

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INTRODUCTION: Basal cell carcinoma (BCC) is the most common skin cancer in humans and accounts for 70-80% of dermatological malignancies. It is known to occur on sun exposed skin. Vulvar and perianal regions are the non-sun exposed atypical sites.[1,2] Vulvar BCC accounts for less than 1% of all BCCs and less than 5% of all vulvar carcinoma.[2]

Vulvar BCC was first described by Temesvary in 1926. Approximately, 300 cases have been reported till date.[3,4,5] Although the aetiology of vulvar BCC is not deciphered, several predisposing factors like chronic irritation and infection, trauma, arsenic, radiotherapy and syphilis[4] have been implicated in the pathogenesis.[5,6] They are slow growing indolent tumours, but are locally invasive and destructive, necessitating adequate surgical excision. They rarely metastasise to inguinal lymph nodes and are known to have local recurrence as high as 20% in some reports,[5,6] which is attributed to inadequate surgical resection.[7]

We present an unusual case of vulvar BCC located on left labium majus in a 50-year-old woman treated with radical local excision.

CASE REPORT: A 50-year-old woman presented to the Dermatology Outpatient Department with complaints of pruritus vulva for past six months. On clinical examination by the Dermatologist, a hyperpigmented plaque was identified on the left labium majus and biopsy was performed as a part of initial evaluation. The punch biopsy was sent for histopathological examination and was reported as basal cell carcinoma. Following this, the patient was referred to Gynaecologic Oncology for further management.

Physical examination revealed a well-demarcated hyperpigmented plaque measuring 2.5 x 1.5 cm on left labium majus (Figure 1). Three satellite lesions, each measuring approximately 0.5 cm were identified surrounding the main lesion. There was no obvious inguinal lymphadenopathy. Rest of the vulva, vagina and cervix appeared normal on colposcopic examination. Pelvic examination revealed no abnormality and conventional cervical Pap smear showed no intraepithelial lesion. As a part of surgical workup, the patient was tested and found to be positive for syphilis which was confirmed by a positive TPHA test.

The patient underwent radical local excision of the tumour which included amputation of the clitoris to get 2 cm surgical clearance and disease free margin. Histopathologic examination of the radical excision specimen showed a 2.5 x 1.5 cm ulceroproliferative growth in the left labium majus. On microscopic examination, nests of basaloid cells were seen arising from the basal layer of epidermis and infiltrating the superficial papillary dermis. These cell clusters showed peripheral palisading and clefting around the tumour nests, a feature characteristic of BCC (Figure 2). The deep dermis was uninvolved and all the surgical margins were free of tumour. The patient is disease free at eight months of postsurgical follow-up.

DISCUSSION: BCC of the vulva is a rare malignancy accounting for less than 1% of all BCCs and less than 5% of all vulvar neoplasm[2]. It usually affects white women over 70 years of age[5]. The relatively younger age at presentation in the present case is noteworthy. Fleury et al has reported a case of vulvar BCC in a 20-year-old patient[7]. All though the aetiology of vulvar basal cell carcinoma is unknown, chronic vulvar irritation, radiation, exposure to arsenicals,

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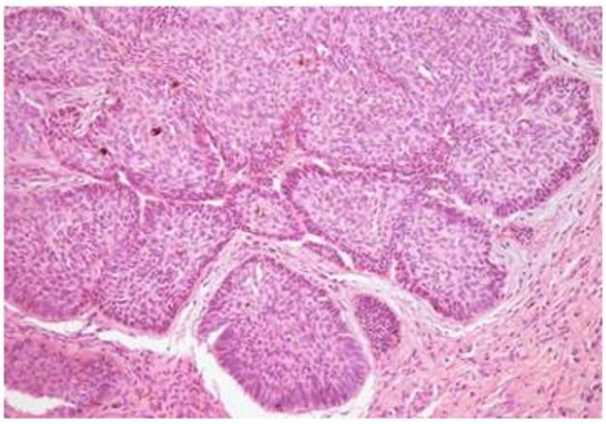
immunodeficiency, syphilis, HPV, nevoid basal cell carcinomas syndrome, xeroderma pigmentosum and mutation in p53 gene are risk factors for vulvar BCC. Our patient was tested positive for syphilis.

Vulvar BCCs usually present as a nodule or as an ulcerated or pigmented lesion[4]. Pruritus (35%), swelling (30%), bleeding (25%) and pain (18%)[1] are the most common presenting symptoms. Our patient had pruritus vulvae with a hyperpigmented plaque. Previous studies demonstrated that the tumour size ranged between 0.2 to 10 cm and most commonly occurred on the labia majora. The less common sites of involvement are labia minora, urethral meatus, prepuce and clitoris.[5,6] In the present report, the tumour was seen involving the left labium majus with clinically suspicious involvement of the clitoris, although clitoral involvement was not demonstrated on histopathological examination.

The diagnosis of BCC of vulva is seldom made clinically. As these lesions are slow growing and indolent, awareness of this entity and a high degree of clinical suspicion is required for early diagnosis. While it could be delayed as long as 20 years,[8] our patient was symptomatic 6 months prior to presenting to the hospital. As vulvar skin is prone to inflammatory and other non-malignant dermatological lesions, BCC is usually misdiagnosed as Bowen's disease, Paget's disease, squamous cell carcinoma (SCC), leucoplakia, lichen planus, and lichen sclerosis. Similarly, in the present scenario initial clinical differentials of SCC, Bowen's disease and Paget's disease were considered and BCC was not suspected.

The conventional treatment recommended for vulvar BCC is wide local excision and adequate clearance with 2 cm margin.[9] High local recurrence rates of 20%[1,2,10] and metastases of 0.1%[10] after simple excision has been noted. Inguinal lymph node dissection at the primary procedure is controversial and is only recommended when the invasion is deep and extensive. Most of the local recurrences have been attributed to inadequate resection.[7] Our patient underwent radical local excision. When surgery is contraindicated or in the case of incomplete excision, radiotherapy is an alternative but often leads to local complications.[6] Postoperative radiation does not appear to reduce recurrence rate or improve overall survival.[7] The role of systemic chemotherapy is limited to metastatic disease. At the postsurgical follow-up of eight months, our patient is free of tumour. However, because of high local recurrence, longterm follow-up is necessary.

Gross Appearance of BCC of Vulva



photomicrograph showing peripheral palisading of the nuclei with retraction from the surrounding stroma (X200 original magnification, HE stain)

CONCLUSION: Any chronic, persistent lesion in vulvar region especially in advanced ages should be subjected to biopsy as BCC has subtle symptoms and nonspecific presentation. Clinician should be vigilant in cutaneous examination as other mucocutaneous diseases like contact dermatitis, psoriasis, Bowen's disease, Paget's are more common on vulva and rare presentation of basal cell carcinomas in genital skin should be kept in mind. It is suggested that all cases of vulvar basal cell carcinomas be registered, treated and followed up in a Gynaecologic Oncology service, irrespective of the status of the margins of surgical specimen. The importance of longterm follow-up has to be highlighted.

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