Pigmented Basal Cell Carcinoma of Vulva: An Unusual Clinico-histomorphological Finding

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Abstract:
Basal cell carcinoma is the most common skin malignancy, often seen in elderly patient with the sun exposed areas, accounting for approximately 70-80% of cutaneous cancer. It can occur in sun protected areas among them genital involvement is very rare and it accounts for less than 1% of all cases. As the vulvar BCC is very rare accounting about less than 5% of all vulvar neoplasm, most of the time it is misdiagnosed due to its nonspecific characteristics. We report a case of vulvar basal cell carcinoma which was misdiagnosed for 2 years in a 39 years old middle-age female patient. Vulvar basal cell carcinoma is usually diagnosed late due to its asymptomatic and slow growth pattern. The cause is unknown, but chronic vulvar irritation is important factor that gives clue to early diagnosis.

Keywords: Vulva, Basal cell carcinoma, Skin, Excision biopsy.

Introduction
Basal cell carcinoma (BCC) is a neoplasm tumor originating from sun exposed skin, generally seen in elderly age group of patient with head and neck involvement predominantly. It quite uncommonly originates from sun-protected skin of genital and perianal region, such as vulva. In literature very few cases have been reported accounting for less than 200 cases. Vulvar BCC generally presents as nodular or ulcerated lesion, accounting for less than 1% of all BCC and less than 5% of all vulvar cancer.

We report a case of Vulvar pigmented nodular BCC in a 39 years old woman. In addition to its uncommon location patient was faultily misdiagnosed and treated for 2 yrs.

Case Report
A 39 year female presented with no prior family history of skin cancer or radiation exposure. She had history of ulcerated growth since two and half yrs. Initially it was tiny nodular, got neglected by patients but later on with irritation and itching sensation got ulcerated for that she had taken treatment from skin specialist who diagnosed as infected dermatitis and treated with topical steroids and antibiotics.

She came to our hospital’s surgical department with history of pruritic non-healing ulcer on left labium major. History narrated by patient that initially lesion was small papular growth, gradually due to chronic pruritus it became a non-healing ulcer. There was no family history of malignancy or skin cancers or any systemic illness and negative for serological test like venereal diseases and HIV. On physical examination growth was 3x2 cm with oval shape, partially hyper pigmented well demarcated noduloulcerative lesion and superficial ulcer of 20mm diameter. Rest of systemic examination and laboratory investigations were absolutely normal. A wide local excision biopsy of growth is performed and sent for histopathological examination. Macroscopic examination of excised growth revealed noduloulcerative lesion with mildly elevated borders measuring 3x2.5 size with the surrounding skin from groin. [Fig-1,2] Microscopically revealed large globules of basaloid cells with peripheral palisading nuclei that project into reticular dermis, stromal retractions, pleomorphism, and cytological atypia along with brownish black pigments in between, typical of pigmented basal cell carcinoma. Lateral and deeper margins were free of tumour invasion. [Fig- 3, 4, 5]

The patient was without any clinical signs of recurrence of growth at 9 months follow up.

Fig. 1: Noduloulcerative lesion with mildly elevated borders.
DISCUSSION

Basal cell carcinoma is the most common skin malignancy in western countries as compared to dark skinned south East Asian countries. Specifically vulvar BCC is rare with only approximately 200 cases reported in literature and seen in less than 1% of all BCC.\textsuperscript{3,4} It is usually seen in sun-exposed areas of skin and relatively seen in sun-protected body parts like genitals, groins, axillae and buttocks.\textsuperscript{5}

In vulvar BCC age group affected ranges from 34 to 96 years, with an average age of 70 years.\textsuperscript{3,6} It might present as nodular or ulcerated or pigmented lesion leading to misdiagnosis of growth as site is extremely rare for its origination. Initially it is asymptomatic but most of the times its presents with itching/pruritus followed by swelling and pain. De Giorgi et al reported that most common symptoms were Pruritus (35%), Swelling (30%), Bleeding (25%) and Pain (18%).\textsuperscript{6} In our case itching/pruritus was initial complaint. It may also have very nonspecific and indolent clinical presentation mimicking other dermatological conditions such as eczema, psoriasis, seborrheic keratosis or angiokeratoma.\textsuperscript{6} Therefore it is recommended to do biopsies of all suspicious vulvar lesion.

Basal cell carcinoma is a multifactorial disease in which both environmental and host factors contributes to its pathogenesis.\textsuperscript{7} Factors other than ultraviolet radiations seems to be involved in etiology of BCC in sun-protected areas. Literatures says that pelvic radiotherapy, chronic pruritus vulvae or ani, vulvovaginitis, history of trauma or burn or certain genetic conditions like nevoid basal cell carcinoma and xeroderma pigmentosa, Human papilloma virus (HPV) infection, p53 gene mutation and advancing age may also contributes to the development of BCC in these sites.\textsuperscript{2,7,8} Similar to our patient, some of those
reported in the literature had no known predisposing factors.

Metastatic BCC is a rare condition with an incidence of 0.0028 to 0.1% and it frequently goes to the inguinal lymph node. It was noted in aggressive histologic pattern like morpheaform, infiltrating and basosquamous type. Metastasis of regional lymph node and perineural invasion were reported. In our case, BCC is diagnosed as Pigmented nodular type, metastasis and lymphatic involvement were not present.

Recommended treatment of choice for BCC is wide local excision or Mohs micrographic excision with pathologically proven clear margins of approximately 1 cm. Continued follow up of the patient is mandatory to check for local recurrence, any evidence of metastasis and any new primary lesion. In our case no recurrence or new growth seen in 9 months strict follow up.

CONCLUSION
This case highlights about the need for vigilance in proper cutaneous examination of any chronic, persistent lesion in the groin keeping in mind about differential diagnosis and rare clinical presentation of BCC in sun-protected areas. It also highlights the fact that young individuals and non-white patients can develop BCC without chronic sun-exposure. Early detection is critical to allow complete wide local excision to prevent further recurrence.

REFERENCES: