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Superficial Basal Cell Carcinoma of the Nipple Mimicking Paget's Disease of the Breast

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ABSTRACT

Basal cell carcinoma (BCC) is a slow-growing, locally invasive form of skin cancer originating from basal cells, and it accounts for 75% of non-melanoma skin cancers. BCCs exhibit various clinical presentations, including nodular, superficial, morpheaform, and others. Most of these tumors appear in sun-exposed areas of the face and neck, while their occurrence in the nipple-areola complex is rare. When BCC arises in these areas, Paget's disease of the breast must be excluded as a possibility. Furthermore, the development of BCCs at a young age, especially with an unusual distribution or multiple lesions, should prompt consideration of genetic conditions such as basal cell nevus syndrome. This case report describes a rare instance of superficial BCC in the nipple-areola complex, initially mistaken for Paget's disease, ultimately leading to the diagnosis of basal cell nevus syndrome.

Keywords: Nipple-areola complex, Basal cell carcinoma, Paget's disease, Basal cell nevus syndrome

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Introduction

Paget's disease of the breast (PDB) is a rare malignancy affecting the nipple-areola complex (NAC), often accompanied by underlying breast cancer [1]. It commonly presents as an itchy, erythematous lesion that may crust or scale, resembling eczema-like conditions. As a result, it is frequently confused with inflammatory skin disorders such as dermatitis or psoriasis, and other tumors, including Bowen's disease, superficial spreading melanoma, and basal cell carcinoma (BCC) [2, 3]. BCC is a slow-growing, locally invasive cancer that originates from basal cells [4]. Although BCC is the most prevalent skin cancer worldwide, its appearance in the NAC is extremely uncommon [5]. The development of BCC is primarily linked to DNA damage from ultraviolet (UV) radiation, and while these cancers generally occur in sun-exposed skin areas, their occurrence in sun-protected regions like the breast is rare, with only around 74 documented cases [6, 7]. Furthermore, BCC can appear sporadically or in conjunction with genetic conditions, such as Gorlin syndrome.

Gorlin syndrome, also referred to as basal cell nevus syndrome (BCNS), is a genetic disorder passed down in an autosomal dominant pattern. It arises from mutations in the PTCH1 gene, resulting in a variety of clinical manifestations that include BCCs, calcified falx cerebri, ovarian fibromas, and odontogenic cysts, which emerge at various ages [8-10]. BCCs are the most frequent manifestation of BCNS, and while they typically develop in sun-exposed regions, they can also occur in sun-protected areas. Clinical diagnostic criteria proposed by Kimonis *et al*. [8] are widely used to diagnose and monitor BCNS. This report presents a case of an elderly woman whose NAC cancer was initially misdiagnosed as PDB, but further examination confirmed it as BCC, ultimately revealing an undiagnosed case of BCNS.

Case report

A 68-year-old woman was referred to dermatology by her gynecologist due to a lesion on her right nipple-areola complex (NAC), which had been present for six months. She had no history of radiation exposure, trauma, or

behaviors that might increase risk. However, she had a history of multiple basal cell carcinoma (BCC) excisions, mostly on the trunk and her face, since her twenties. On physical examination, a 3 cm, clearly defined, red, ring-shaped plaque with an ulcerated surface was found, covering the entire right NAC **(Figure 1)**. There was no evidence of axillary lymph node involvement, and the left breast appeared normal on both inspection and palpation.

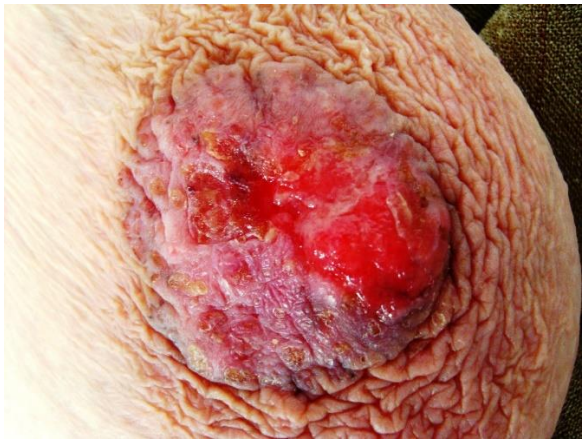

Figure 1. Basal cell carcinoma of the right nipple-areola complex showing ulceration.

Upon further examination, additional findings included palmar pits **(Figure 2)**, multiple milia on the fingers and face, and an epidermal cyst on the left thumb. Aside from scars from prior surgeries on her face and trunk, the rest of the physical test was unremarkable. Paget's disease of the breast (PDB) was initially suspected, but mammography showed no signs of breast malignancy, and there was no enlargement of axillary lymph nodes on the ultrasound.

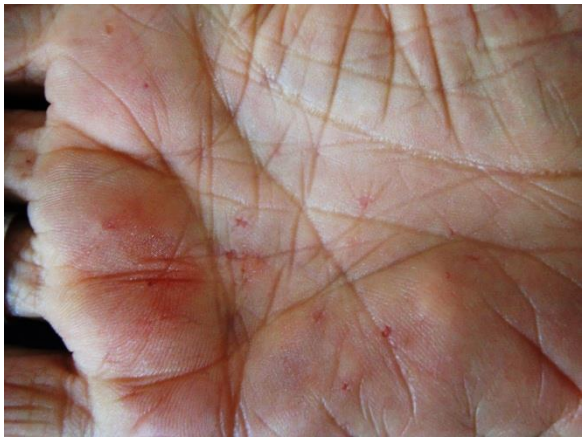

Figure 2. Multiple palmar pits.

The histopathological examination of the biopsy from the lesion showed small clusters of abnormal basaloid cells, well-defined and located in the lower part of the epidermis. These clusters were contained within the papillary dermis, with no signs of spreading. Based on these findings, a diagnosis of superficial basal cell carcinoma (BCC) was made. Consequently, the lesion was surgically removed. The clinical presentation and histopathological results fulfilled the diagnostic criteria for basal cell nevus syndrome (BCNS), which includes two primary criteria: the occurrence of multiple BCCs since adolescence and the presence of more than three palmar pits. No other significant features of BCNS were identified in the physical or imaging exams. The patient has been monitored for approximately three years, with no signs of recurrence or further issues.

Results and Discussion

Non-melanoma skin cancers are among the most prevalent cancers in individuals with fair skin, with BCC accounting for around 75% of all cases. These cancers typically develop in areas of the skin exposed to the sun, like the head and neck, though in rare instances, they can appear in areas not commonly exposed to sunlight, including the nipple-areola complex (NAC) [11, 12]. A review by Chun and Cohen [13] identified 55 cases of BCC in the NAC, with 19 more reported in subsequent studies. From these cases, two key trends can be observed. First, BCCs in the NAC are more common in men, possibly because males are more likely to expose their chest to sunlight compared to females. Some experts suggest that UV radiation could still be a contributing factor, even in these rare NAC cases. Second, BCCs in this area tend to have a more aggressive nature than those found in other parts of the body, such as the head or neck. This increased aggression may be linked to a higher rate of metastasis, estimated at 4% (3 out of 74 cases), although some studies have disputed this conclusion [14-16].

Basal cell carcinoma (BCC) presents in various forms, with the most common subtypes being nodular, micronodular, superficial, morpheaform, infiltrative, and fibroepithelioma of Pinkus. BCCs occurring in the nipple-areola complex (NAC) are more likely to be the nodular subtype (42.9%), while superficial BCCs, like the one in our case, account for less than 31% of such instances in this region. Clinically, superficial BCC appears as a red macule or thin plaque with poorly defined edges, often with a scaly or crusted surface. This can lead to misdiagnosis as other neoplastic conditions like Bowen's disease or Paget's disease of the breast (PDB), as well as benign skin conditions such as dermatitis [15-18]. In our patient's case, PDB was initially suspected but was later excluded following histological examination.

Though the majority of BCC cases are sporadic, genetic disorders such as basal cell nevus syndrome (BCNS) should be considered in patients with multiple BCCs, especially those developing from a young age or with an unusual distribution. BCNS is a rare autosomal dominant multisystem disorder that exhibits full penetrance but variable expression. The main features of BCNS include BCCs, palmar or plantar pits, odontogenic keratocysts, and intracranial calcifications. To diagnose this syndrome, patients must meet two major criteria or one major and two minor criteria [16-19]. Our patient met two major criteria: the presence of multiple BCCs with early onset and more than three palmoplantar pits.

Conclusion

This case of superficial BCC is unique due to its occurrence in an uncommon location (the NAC), its association with BCNS, and its clinical resemblance to PDB. To our knowledge, this is the first reported case of BCC in the NAC within the context of BCNS.

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Ethics Statement: None

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