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Direttore Nicola Picardi

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Report of a case and literature review

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Emilio Trignano, Matteo Jacopo Manzo, Panagiotis Paliogiannis, Massimo Madonia, Francesco Marongiu, Amelia Lissia, Gian Vittorio Campus

Department of Surgical, Microsurgical and Medical Sciences, University of Sassari, Sassari, Italy

Basal cell carcinoma of the nipple-areola complex. Report of a case and literature review

Basal cell carcinoma of the nipple areola complex in female patients is an extremely rare tumor with a malignant clinical behavior.
The Authors present the case of a Basal cell carcinoma of the nipple areola complex in a Caucasian woman, evaluated by clinical and histological findings.
Our clinical experience suggests that Basal cell carcinoma of the nipple areola complex has a local aggressive behavior with an higher metastatic potentiality compared to basal cell carcinoma of common sites. However, an accurate tissue sparing excision may be successful in the long term.

KEY WORDS: Basal cell carcinoma, Nipple areola complex, Tissue sparing excision

Introduction

Basal cell carcinoma of the nipple areola complex (NAC Bcc) is an uncommon tumor. NAC Bcc is a biologically malignant neoplasm, with a higher metastatic potentiality compared to Basal cell carcinoma (Bcc) of common sites [1].
To date, only 13 cases of NAC Bcc in female patients have been reported in English literature.
The present study describes the history of a 40 years old woman with a NAC Bcc of the right nipple. Clinical and histological findings are discussed along with differential diagnosis, therapy and outcomes.

Case presentation

A 40 years old Caucasian woman presented to our department with a 10 months history of a growing lesion on her right nipple areola complex. The patient had noticed a progressive increase in size of the lesion, with pruritus and weeping as symptoms. Physical examination revealed a 0,7cm cutaneous nodular ulcerated lesion involving the right nipple areola complex (Fig. 1). No axillary lymphadenopathy was found. The patient had no previous or family history of skin cancer and past medical history was not significant. She also had no history of sun, radiation or arsenic exposure.
According to the clinical findings and after a consultation with dermatologists, a clinical suspected of Bcc came

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Correspondence to: Dr. Panagiotis Paliogiannis, Department of Surgical, Microsurgical and Medical Sciences, University of Sassari, Viale San Pietro 43B, 07100, Italy (e-mail: ppaliogiannis@uniss.it)

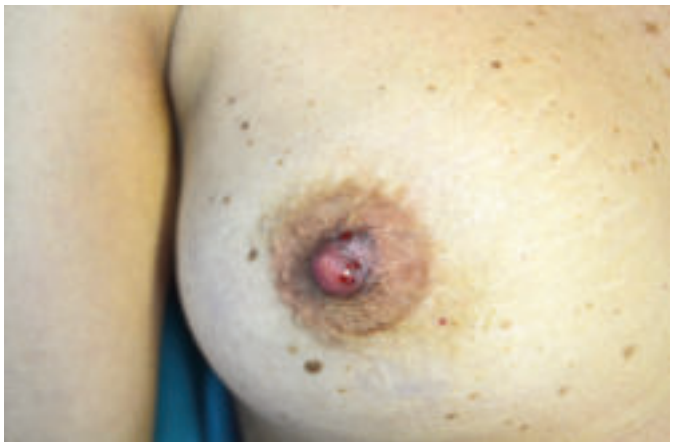

Fig. 1: Preoperative frontal view of the right nipple-areola complex.

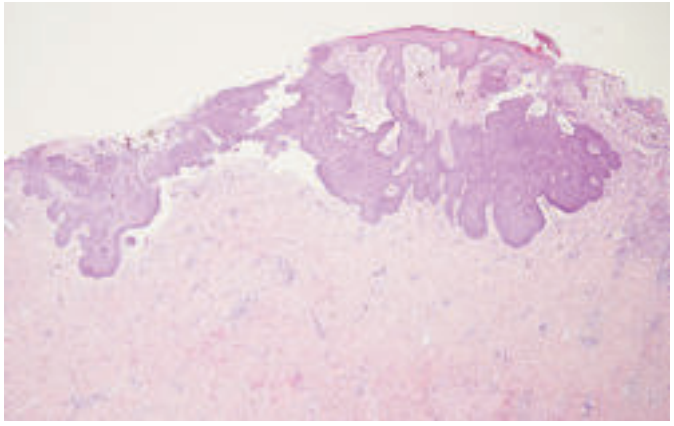

Fig. 2: Haematoxylin-eosin stained section of the neoplasm.

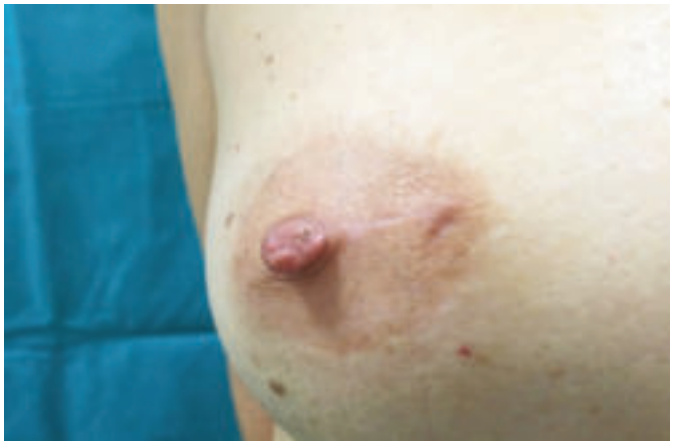

Fig. 3: Postoperative frontal view of the right nipple-areola complex.

up and surgery was planned. A tissue sparing skin excision, including 2mm macroscopic safety margin, was performed under local anaesthesia. A dressing with a soft, low friction top film was used, as we previously reported[2].

The pathologic findings confirmed the clinical suspect: ulcerated nodular Bcc with well circumscribed basalioid tumor lobules with no milk ductal infiltration and neoplasm free margin on all specimens (Fig. 2).
The patient was successfully followed up for 18 months and no local relapse or systemic disease has been registered (Fig. 3).

Discussion

Bcc is the most common skin human malignancy in the western world, affecting routinely sunlight exposed regions[3]. Like other malignancies it rarely involves the nipple[4]. Indeed, NAC Bcc is an extremely uncommon neoplasm. From 1893 to date only thirty-nine cases have been described in English literature and only thirteen in female patients, nine of these described in literature since 2000. This could be explained by the changing in clothing and sunlight exposure habits in last years.
Analyzing literature data appears that NAC Bcc has a greater ability in lymphatic metastasis compared to Bcc of common sites. This could be motivated by the presence of a rich lymphatic network in the sub-areolar plexus, that may provide a direct way for tumor spread [4].
Bcc recognizes several risk factors, the most significant of which are ultraviolet light and radiation exposure. Others are previous history, family history, trauma, arsenic exposure, immunocompromised status, prior burns, basal cell nevus (Gorlin's syndrome). However, medical records of NAC Bcc female patients do not allow us to identify a clear risk factor in this small subgroup.
Given the wide range of morphology, NAC Bcc can potentially be mistaken for other benign and malignant skin lesions, such as Paget disease, Bowen syndrome, eczema, erosive adenomatosis and malignant melanoma. Most women with a histopathological diagnosis of Paget disease present a clinical abnormality of the nipple. However, in at least 10% of affected patients, Paget disease is found incidentally, during microscopic examination of mastectomy specimens [5]; the differential diagnosis is generally based on histopathological and immunohistochemical analysis.
Congdon and Dockerty in 1956 reviewed malignant breast disease from Mayo clinic since 1910 and firstly described a case of a NAC Bcc in a female patient [6].
Farrow in 1958 and Davis in 1977 presented the cases of a 49 and 66 years old women with NAC Bcc, treated with biopsy, followed by wide local excision [7,8].
Sauven in 1983 reported the case of NAC Bcc in a 75 years old female patient, suggesting a limited wedge biopsy and subsequent radiotherapy as treatment [9].
More recently, Sánchez-Carpintero in 2000 reported the case of a 75 years old patient with a 20-years history of a wide nipple lesion extending into the lactiferous duct and first described the use of Mohs' micrographic surgery

in NAC Bcc patients [10]. This technique was also used by Zhu et al. in a 47 years old woman [11].
Yamamoto and colleagues in 2001 described the case of a 82 years old female patient with a NAC Bcc with intraductal extension [12]. After an incisional biopsy, the lesion was resected with a 2 cm free margin along with the underlying mammary tissue [13]. Betti et al. in 2003 reported the case of a 47 years old woman underwent NAC Bcc excision. From their point of view, literature reports do not suggest that these BCCs have an increased potential for malignancy [14].
Huang and colleagues in 2005 describe their experience with a NAC Bcc in a 46 years old woman. After an incomplete excision, a partial mastectomy with excision of the sentinel lymph node was performed [15]. Rosen in 2005 used Mohs' surgery and sentinel lymph node biopsy (resulted negative for tumoral invasion) in a NAC Bcc of a 49 years old female [16]. We believe that the sentinel lymph node biopsy does not have a valid meaning without clinical and/or radiological suspicion.
Sharma and colleagues, in 2011, reported a case of a wide pigmented NAC Bcc in a 48 years old female. After a needle biopsy resulted as inconclusive, a two cm margin excision was performed [17]. Jung in 2011 described the history of an asymptomatic NAC Bcc presented as black plaque on the areola of the right breast present for about 30 years in a 67 years old woman. After a size increasing in last time, the tumor, including a 5 mm safe margins, was excised [18]. Xu et al. in 2012 reported a case of a 72 years old chinese female with the presance of an erosive lesion excised with 3 cm surgical margins [19].
From these data appears how variables are the age, the speed of onset and the way of presentation. Varying modes of treatment have been described ranging from medical treatment, Mohs microsurgery, local excision with or without radiotherapy, partial mastectomy with axillary sentinel node dissection.
In our opinion, wide excision may be avoided, as confirmed by the fact that, in most cases, Mohs microsurgery and simple excision was not followed by recurrence. Considering the importance of this region in women and in order to improve the aesthetic outcome, our experience suggests that tissue sparing excision, with 2mm macroscopic safety margin, could be the first choice of treatment in these patients. A routine follow-up of at least 5 years should be suggested.

Riassunto

Il carcinoma basocellulare del complesso areola-capezzolo femminile è una neoplasia estremamente rara, caratterizzata da un comportamento clinico maligno. Presentiamo in questo articolo un caso di carcinoma basocellulare del complesso areola-capezzolo in una donna caucasica, con particolare riferimento agli aspetti clinici ed istopatologici. La nostra esperienza suggerisce che la neoplasia ha un comportamento aggressivo tanto localmente quanto in riferimento al suo potenziale di metastatizzare a distanza, che si rileva superiore a quello dei carcinomi basocellulari di altre sedi. Ciononostante, un'accurata escissione "tissue sparing" può garantire buoni risultati a lungo termine.

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