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Primary Axillary Tumors: About Two Cases

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Abstract

Cutaneous carcinomas are mainly of keratinocyte or adnexal glandular origin [1]. Cutaneous primary mucinous carcinoma is a rare adnexal tumor developed from the sweat glands with low metastatic potential, but skin recurrences are common [2]. Eccrine porocarcinoma (EPC) is a rare and aggressive skin cancer, accounting for approximately 0.01% of skin tumors [3,4]. It can manifest as papules, plaques or nodules and can appear on any part of the body, although it usually affects the head, neck and lower extremities. CPE is often misdiagnosed as squamous cell carcinoma or Bowen's disease, highlighting the importance of the exam.

We report the cases of two patients operated on for primary axillary tumors. The first case was a tumor recurrence after two previous excision gestures and where the assessment did not objectify another primary focus of mucinous carcinoma. The second case was a large axillary tumor suggestive of metastatic breast cancer (lymph node cancer), radiological investigations and immunohistochemical study was in favor of a primary porocarcinoma of the skin. In both cases we performed an enlarged resection with lymph node dissection.

Keywords

Cutaneous Primary Carcinoma, Axillary Tumors, Mucinous Carcinoma, Eccrine Porocarcinoma.

Introduction

Axillary masses pose a diagnostic challenge as several benign and malignant entities, primary and secondary, are potential causes.

The search for another potential primary tumor is an important step in the diagnostic process. Differential diagnosis is sometimes difficult with breast cancer in its lymph node form, and morphological and radiological assessment are essential to eliminate a lymph node form of breast carcinoma. The axillary region can also be the preferred site of certain digestive, bone and other cancers, as such the search for a possible primary location is part of the pretherapeutic assessment of an axillary tumor. The definite diagnosis of primary cancers is made by immunohistochemistry. Surgery remains the essential treatment.

Adnexal carcinomas are rare skin tumors derived from eccrine and apocrine sweat glands, hair follicles and sebaceous glands.

Among carcinomas derived from the sweat glands, porocarcinoma (EPC) is known for its high rates of recurrence and metastasis, with 20% of cases having metastases, mainly to regional lymph nodes [5, 6].

On the other hand, mucinous carcinoma is generally of good prognosis and the occurrence of metastases is rare. These tumors are characterized by a high rate of recurrence.

Proper surgical resection can achieve curative results in 70% to 80% of cases. In cases of metastatic or recurrent disease, chemotherapy and/or radiotherapy may be added, although there is no consensus on the specific chemotherapy regimen [5, 7, 8, 9] due to the rarity of the disease.

In the following sections, we present the cases of two patients with axillary EPC.

Observation 1

The patient is a 41-year-old man, with no notable medical history, who presented for a recurrent left axillary mass. The history of his illness seems to date back four years ago and is marked by the occurrence of a painless left axillary swelling, for which the patient underwent two excision gestures a few months apart. This swelling reappeared 18 months ago with a gradual increase in size, an ultrasound of the soft parts was done having objectified a heterogeneous, oval and well limited hypoechoic mass (30x10mm), without proper wall with posterior strengthening and respect for the muscular plan. Neglected by the patient, the mass continued

to increase in size. We also found a hard mass of about 6cm of major axis at the level of the left axillary region, irregular and not very mobile, surmounted by a cauliflower keloid scar (Figure1a). Ultrasound of the soft parts revealed an axillary mass syndrome in contact with the axillary vascular space measuring on average (55x32mm). The clinical assessment and additional examinations did not find any primary tumors or metastatic sites. Breast and prostate examination were normal. Prostate-specific antigen (PSA) testing, serum carcinoembryonic antigen (CEA) and Ca19.9 levels were within normal range. Ultrasound mammography did not reveal abnormalities, thoraco-abdomino-pelvic CT (TAP CT) and magnetic resonance imaging (MRI) did not objectify other primary or secondary tumor sites, nor bone scintigraphy had objectified arguments that could evoke secondary bone locations.

It was a cutaneous localization of a primary mucinous carcinoma. A wide excision of the lesion with safety margin was performed, associated with a lymph node dissection in front of the presence of multiple lymphadenopathies in the three relays, there was no lymphadenopathy in the axillary hollow (Figure: b, c).

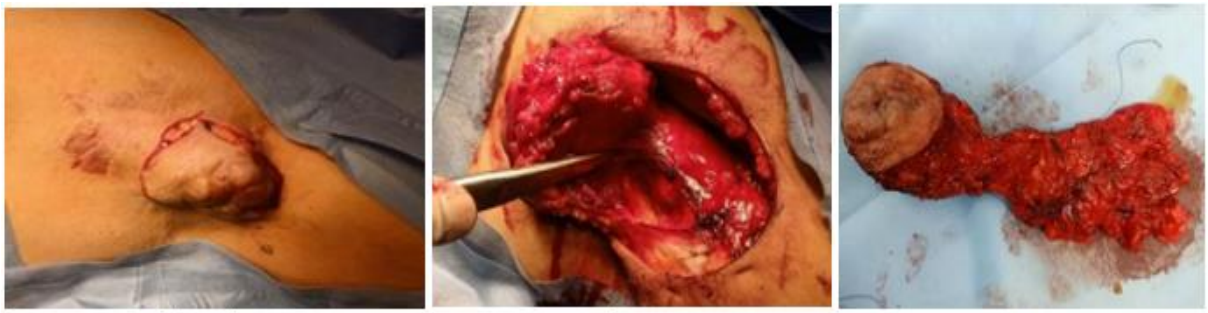

Figure 1: a. Macroscopic appearance of a primary mucinous carcinoma; b. Axillary mass with lymph node dissection; c. Macroscopic appearance.

Histopathological analysis objectified a skin coating whose dermis is the site of an infiltrative carcinomatous tumor proliferation, building vast areas of mucoid substance in which glandular structures and isolated tumor cells were found. Cytokeratin immunostaining (CK) revealed a CK7+/CK20− /p63−/Rp−/Ro− profile.

In conclusion, the histological appearance responded to a cutaneous localization of a mucinous carcinoma most likely of primary origin with the presence of axillary lymph node metastases (20N + / 30N), the limit of upper surgical excision was healthy, located 3mm from the tumor. (Figure 2: a, b).

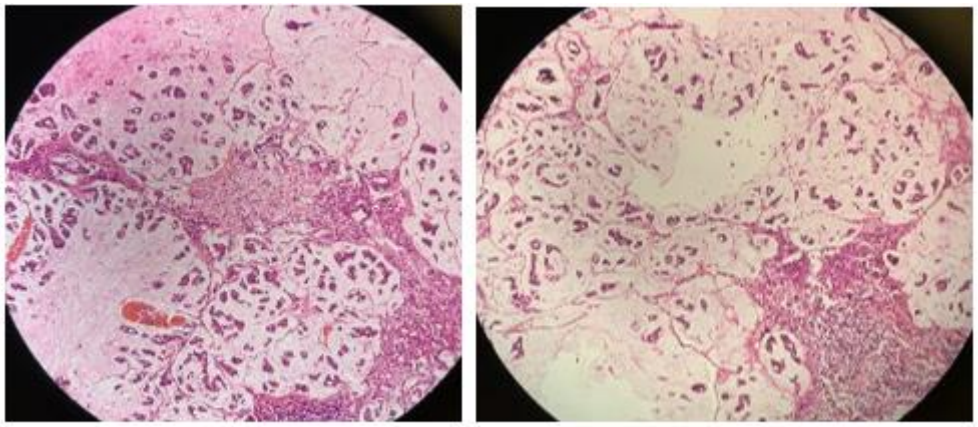

Figure 2 (a, b): Glandular structures surrounded by abundant extracellular mucin.

Observation 2

The patient was a 62-year-old man with no significant medical history except for benign prostatic hyperplasia under treatment. He had three axillary swellings that were gradually increasing in size.

Axillary ultrasound revealed the presence of three hypoechoic right axillary lymph nodes, measuring 26 x 18 mm and 13 x 7 mm, and the largest had irregular hypoechoic areas measuring 63 x 43 mm. computed tomography (CT) scans showed several axillary right lymph nodes, the largest measuring 19x07 mm. Initially treated as a probable lymph node metastasis of breast carcinoma (based on biopsy results and immunohistochemical profile: IHC: ER, negative, PR negative, HER2 negative and KI67.50%). No detectable secondary locations to bone scanning. A biopsy part re-evaluation revealed a histopathological aspect consistent with lymph node metastasis of poorly differentiated carcinoma, the primary origin remains to be determined, IHC profile (CK7 Positive, CK20 negative, PR negative, RO negative, PS100 negative, chromogranin negative, and TTF1 negative). The patient received 8 cycles of chemotherapy consisting of Cyclophosphamide, Taxotere and Adriamycin.

On admission, the clinical examination showed no fever and no signs of general aggravation, apart from a weight loss of 5 kg. Three painless, motile, and ulcerated axillary swellings were observed. It should be noted that the tumor volume decreased after chemotherapy. Tumour markers were negative (CA 15-3: 21.40 IU/ml, CA 19-9: 37.56 IU/ml, CEA: 6.154 ng/ml).

Treatment consisted of block excision of the tumour with axillary lymph node dissection of all three levels of Berg, as well as a total mastectomy of the right breast. (Figure 3).

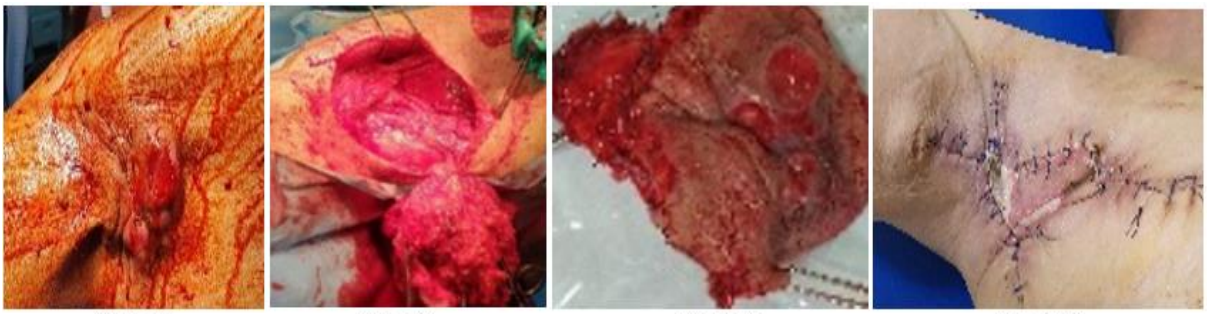

Figure 3a Figure 3b Figure 3c Figure 3d

Figure 3: a. Macroscopic appearance of primary eccrine porocarcinoma; b. Surgical procedure; c. Resection piece; d. Parietal plasty.

Histological and immunohistochemical results suggestive of porocarcinoma of the skin, presence of vascular and tumor emboli and lymphatic infiltration 12 lymph nodes infiltrated on the 25 lymph nodes removed (12 N^+ / 25N). On the other hand, the histopathological study of the mastectomy piece did not objectify lesions or tumor infiltration (figure 4).

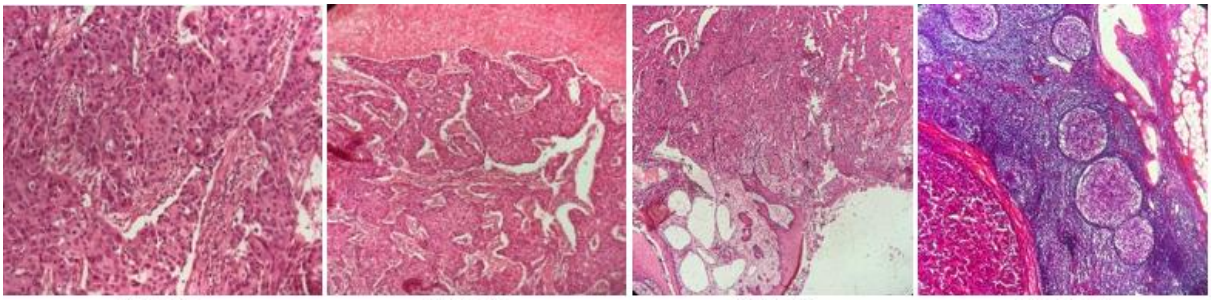

Figure 4a Figure 4b Figure 4c Figure 4d

Figure 4: a. Atypical tumors cells; b. Tumor necrosis and tumor proliferation; c. Tumoral proliferation and ulceration; d. Lymph Node Metastasis.

Discussion

Eccrine sweat porocarcinoma (ESP) is a malignant skin tumor that develops from the eccrine sweat glands, while apocrine tumors are particularly located at the axillary, inguinal and anogenital level [10, 11]. These tumors occur mainly around the age of 60, but there are extremes from 8 to 84 years [2, 12, 13], affects both sexes with a male predominance (sex ratio of 2:1) [2, 13].

Primary cutaneous mucinous carcinoma is a very rare adnexal malignancy developed at the expense of the sweat glands, representing 0.005% of all skin tumors [2], first described by Lennox et al. [12]. It is located mainly in the cervicofacial region in 80% of cases [7, 10], particularly palpebral in nearly 40% [2], but it also affects the trunk and limbs. It is a dermohypodermic tumor, characterized by low-grade malignancy and slow growth [1, 2]. At immunohistochemistry, this tumor expresses low molecular weight cytokeratin, ACE and S100 protein, with negative cytokeratin 20, thus eliminating a digestive origin [14]. Although it has a low metastatic potential, the rate of local recurrence remains high, sometimes late [15]. This tumor can simulate a metastasis of mammary or digestive origin.

The tumour usually takes on a benign but deceptive clinical picture of a solitary, asymmetrical, nodular, irregular flesh-coloured, painless, medium-sized 2 cm lesion with a slow-growing, non-aggressive development [10, 14]. Rapidly progressive forms were observed [14, 15]. It can only be diagnosed by histopathological and immunohistochemical examination and by excluding a primary or metastatic tumor [15].

Eccrine porocarcinoma is an extremely rare malignancy that starts in the intradermal part of the sweat gland. It was first described by Pinkus and Mehregan in 1963 [16], and later the term "eccrine porocarcinoma" was introduced by Mishima and Moriko in 1969 [17]. Porocarcinoma occurs equally in both sexes and is more common in the elderly. It usually presents as an ulcerated nodule or plaque, commonly found in the lower extremities (50%), trunk (24%) or head and neck (24%). The prognosis varies, and a large local excision allows curative results. However, lymph node metastases and distant metastases are associated with a poorer prognosis. It has been reported that this cancer can develop from an eccrine porome. Bone involvement is rare in this tumor and, in our case, the bone was not affected.

There are several architectural histological forms of eccrine porocarcinoma. The in-situ form, which has a good prognosis, resembles eccrine porome but has cytonuclear atypia without invasion. Infiltrating forms include a well-defined trabecular variant with a better prognosis and a poorly defined multifocal epidermotropic variant with a poorer prognosis, associated with a high risk of recurrence and metastasis.

Microscopic examination usually reveals atypical tumor cells arranged in cords and lobules, affecting both the dermis and the epidermis. Tumor cells have nuclear atypia, frequent mitosis and necrosis. Robson et al. reported several histopathological factors predictive of poorer clinical outcomes and death: (1) presence of more than 14 mitosis per high-magnification field, (2) lymphovascular invasion, (3) tumor depth greater than 7 mm, and (4) infiltrating margins [18].

The treatment of choice is broad excision surgery. Surgery alone showed curative results in 70% of cases, although a recurrence rate of 20% was observed. Regional lymph nodes should be evaluated, as porocarcinoma tends to invade the dermal lymphatics, resulting in lymph node involvement in about 20% of cases [19]. In our patient, a wide local excision was performed at the same time as a lymph node dissection due to lymph node involvement.

Chemotherapy is primarily reserved for cases of metastatic disease, although the response rate remains uncertain [20]. Primary surgery offers the best results for localized eccrine porocarcinoma. Given the high probability of lymph node involvement, evaluation of regional lymph nodes, especially in patients with large tumor size (greater than 5 cm), is recommended. The use of adjuvant radiotherapy may be considered, especially in patients at high risk of local recurrence. Adjuvant radiotherapy should be evaluated on a case-by-case basis, especially in patients with close surgical margins and extracapsular extension [21].

Conclusion

The development of adnexal carcinomas in the axillary region poses a diagnostic problem because they can simulate metastases of other primary tumors mainly mammary carcinomas, so it is imperative to make a systematic search for a primary tumor other than that of the skin. Several histological types have been described depending on the point of departure, mucinous carcinoma and procarcinoma derived from the sweat glands, can have the same macroscopic appearance, however in both, the diagnosis can only be affirmed by immunohistochemistry.

Mucinous carcinoma is characterized by slow progression and low risk of metastasis; however, recurrences are frequent. Surgical resection is sufficient in the majority of cases. Porocarcinoma is characterized by an evolutionary risk, essentially the development of lymph node and visceral metastases. Porocarcinoma can be cured by extensive excision, while the metastatic phase is largely insensitive to treatments.

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