Eccrine Porocarcinoma with Squamous Differentiation in a Patient with Oculocutaneous Albinism

Abstract

Eccrine porocarcinoma is a rare malignant skin appendage tumor of sweat gland origin. Eccrine porocarcinoma arising in a patient of oculocutaneous albinism is extremely rare and only two cases have been reported in English literature to the best of our knowledge. Out of the two cases of eccrine porocarcinoma in oculocutaneous albinism, one case had squamous differentiation. We report a case of eccrine porocarcinoma with squamous differentiation in a 39-year-old male, who presented with a nodular lesion on the upper left chest wall. He also had nodal and distant cutaneous metastasis.

Keywords: *Albinism, eccrine porocarcinoma, squamous*

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Introduction

Albinism is a genetically inherited disorder, characterized by a lack of melanin pigment production.[1] Albinism is broadly divided into two types: ocular albinism and oculocutaneous albinism.[1] Oculocutaneous albinism affects the eyes, skin, and hair while the ocular type affects the eyes solely.[1] The oculocutaneous type is an autosomal recessive disorder, and the ocular variant is a sex-linked disorder.[1] Albinos are at an increased risk of developing skin malignancies due to melanin pigment deficiency, which predisposes to harmful effects ultraviolet radiation.[1] The most common cutaneous malignancy reported in albinos is squamous cell carcinoma.[1] The other malignancies reported in albinos are basal cell carcinoma, basosquamous carcinoma, and malignant melanoma.[1,2] However, eccrine porocarcinoma arising in a setting of oculocutaneous albinism is extremely rare.[3,4]

Case Report

A 39-year-old male presented with a nodular lesion on the upper left chest wall for the last 6 months' duration [Figure 1]. The lesion measured (3.5 × 3 × 1) cm and was mildly tender. The patient had oculocutaneous albinism. Family history revealed that the patient had one younger brother. His parents had consanguineous marriage. None of them are affected by albinism.

On general examination, he had pallor with loss of weight. On systemic examination, he had left palpable axillary lymph node. He had an ulcer measuring (2 × 1) cm over the shin of the left leg, which appeared for the last 1 month.

Laboratory investigations revealed anemia with hemoglobin of 8.2 g/dL. The antiretroviral serology was negative. An incisional biopsy from the nodular lesion over the chest wall revealed an infiltrating tumor in the dermis composed of polygonal to cuboidal cells arranged in lobules, nests, and cords which are separated by thick fibrous septa [Figure 2a]. These cells had vesicular nuclei with prominent nucleoli and moderate to abundant pale cytoplasm [Figure 2b]. An intraepidermal component composed of nests and islands of small basaloid cells, sharply demarcated from the adjacent keratinocytes was present. Areas of focal clear cells and malignant squamous differentiation, with keratin pearl formation, were seen [Figure 2c]. The tumor had duct formation and mitoses up to 16 per high-power field. The clear cells were periodic-acid-Schiff (PAS) stain positive and diastase labile. On immunohistochemistry, the tumor cells were diffusely positive for cytokeratin AE1/AE3 [Figure 2d]. Epithelial membrane antigen (EMA) and

How to cite this article: Dey B, Raphael V, Harris C. Eccrine porocarcinoma with squamous differentiation in a patient with oculocutaneous albinism. Indian Dermatol Online J 2020;11:398-400.

Received: 02-Jun-2019. **Revised:** 22-Aug-2019. **Accepted:** 01-Jan-2020. **Published:** 10-May-2020.

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Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ_223_19

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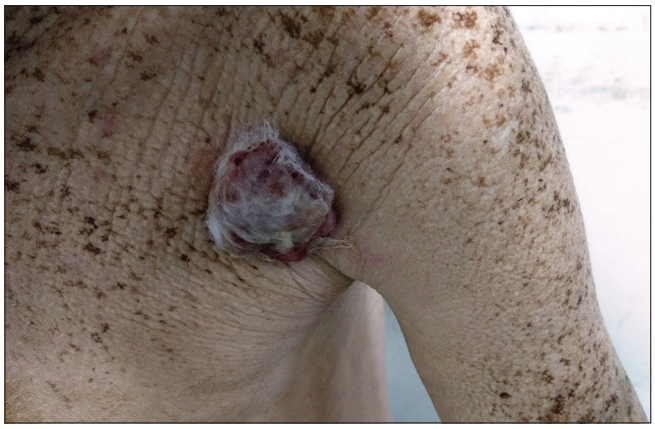

Figure 1: Nodular lesion over the left upper chest wall

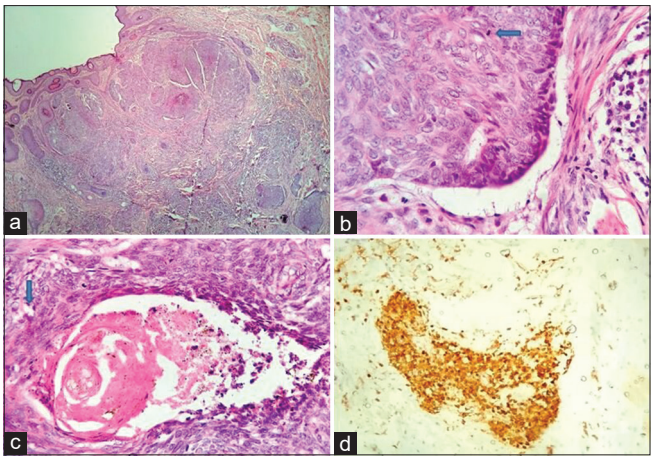

Figure 2: (a) Infiltrating tumor in the dermis (H and E, 4×); (b) tumor composed of polygonal to cuboidal cells arranged in nests separated by thick fibrous septa. Mitosis marked by arrow (H and E, 40×); (c) areas of squamous differentiation. Duct formation marked by an arrow (H and E, 40×); (d) tumor cells positive for cytokeratin AE1/AE3 (immunohistochemistry, 10×)

carcinoembryonic antigen (CEA) highlighted the ducts. A diagnosis of eccrine porocarcinoma with squamous differentiation was made. Fine-needle aspiration cytology from the left axillary node revealed scattered tumor cells in a lymphoid background compatible with metastasis from the chest wall eccrine porocarcinoma with squamous differentiation. A skin biopsy from the ulcer edge over the left leg revealed a tumor morphologically similar to the tumor over the chest wall suggestive of cutaneous metastasis.

A wide excision of the chest wall tumor with axillary lymph node dissection was planned. In view of the distant cutaneous metastasis, adjuvant chemoradiation was planned. However, the patient refused any further treatment and was lost to follow-up.

Discussion

Cutaneous skin tumors are known to occur in albinos. Eccrine porocarcinoma arising in a patient of oculocutaneous albinism is rare and only two cases have been reported in English literature to the best of our knowledge.[3,4] First is a case of eccrine porocarcinoma on the forehead in a 28-year-old oculocutaneous albino male.[3] The patient had left submandibular gland metastasis with no other associated lesion or disease.[3] The other is a case of eccrine porocarcinoma with squamous differentiation on the right cheek in a retro-positive 33-year-old oculocutaneous albino female.[4] The patient had lymph node metastasis with multiple associated lesions such as basal cell carcinoma, squamous cell carcinoma, and actinic keratosis.[4]

Eccrine porocarcinoma is a rare malignant skin appendage tumor of sweat gland origin.[4] The average age of onset is 68 years with no gender predilection.[4] The common sites of occurrence are lower extremities followed by trunk, head, and upper extremities.[4] The etiology of eccrine porocarcinoma is not known. Eccrine porocarcinoma has been reported in two patients of oculocutaneous albinism and in a patient with xeroderma pigmentosum.[3-5] Moreover, a subset of eccrine poroma has been described in actinic keratosis.[6] Based on these observations, it has been contended that ultraviolet radiation may be involved in the genesis of eccrine porocarcinoma especially in those who are genetically predisposed to sun-induced skin cancers.[4]

Squamous differentiation, clear cell areas, focal melanin pigment, and necrosis are not uncommon in eccrine porocarcinoma.[5] Out of the two cases of eccrine porocarcinoma in oculocutaneous albinism reported in the literature, one case had squamous differentiation associated with human immunodeficiency virus (HIV) positivity.[4] Moreover, a subgroup of eccrine porocarcinoma with squamous differentiation has been associated with conditions such as actinic keratosis and Bowen's disease, which are also known risk factors for the development of cutaneous squamous cell carcinoma.[4] Thus, the histogenesis of squamous differentiation in a subset of eccrine porocarcinoma may be attributed to ultraviolet radiation and chronic immunosuppression.[4] In the present case, it may be attributed to ultraviolet radiation in an already genetically predisposed patient of oculocutaneous albinism.

The important differential diagnosis of eccrine porocarcinoma with squamous differentiation is cutaneous squamous cell carcinoma. Porocarcinoma can display prominent squamous differentiation and be mistaken for a squamous cell carcinoma.[4,7] Likewise, squamous cell carcinoma may be erroneously diagnosed as porocarcinoma if it shows pagetoid intraepidermal spread, colonizes the adnexal structures, and shows acantholysis with pseudogland formation. Histopathologically, ductal differentiation and presence of intracytoplasmic lumina help in its distinction from cutaneous squamous cell carcinoma.[4,7] Immunostains may be helpful for difficult cases. On immunohistochemistry, tumor cells in eccrine porocarcinoma are diffusely

positive for cytokeratins (34BE12, AE1/AE3) and p63.[7] CAM 5.2 antigens, CK7, EMA, and CEA highlight the ducts.[7] Positivity for S100 is seen in the scattered dendritic melanocytes.[7]

Prognostically, eccrine porocarcinoma has a poorer prognosis as compared to that of cutaneous squamous cell carcinoma.[7] Eccrine porocarcinoma tends to have increased local recurrence (20%), nodal (20%), and systemic metastasis (11%).[7] Systemic metastasis to lungs, pleura, brain, mediastinum, retroperitoneum, and breast are reported.[8] Distant cutaneous metastasis is also reported.[7] Patients with lymph node metastasis have a mortality rate as high as 67%.[4] Poor prognostic indicators on post-excision histopathology include high mitosis of more than 14 per high-power field, lymphovascular invasion and depth of tumor more than 7 mm.[9] In the present case, the patient had both nodal and distant cutaneous metastases. The patient also had a high mitotic rate of 16 per high-power field.

The standard treatment is wide local excision with histologically clear margins.[7] Few reports have also described the benefits of radiation and chemotherapy.[7] The optimal treatment has not been standardized yet as there is little evidence of a specific chemotherapy-based regimen in metastatic eccrine porocarcinoma due to its rarity.[8] Varieties of chemotherapeutic agents used in combination are doxorubicin, mitomycin, vincristine, and 5-fluorouracil; anthracycline, cyclophosphamide, vincristine, and bleomycin; isotretinoin and interferon-alpha; carboplatin and paclitaxel; cisplatin and docetaxel; paclitaxel and interferon-alpha. Each of these combinations has variable response rates and toxicities.[8]

Eccrine porocarcinoma arising in a patient of oculocutaneous albinism is extremely rare. Histogenesis of squamous differentiation in a subset of eccrine porocarcinoma may be attributed to ultraviolet radiation and chronic immunosuppression. It has a poor prognosis with aggressive behavior.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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