Journal of Clinical & Experimental Dermatology Research

Abduljabbar et al., J Clin Exp Dermatol Res 2016, 7:5
DOI: 10.4172/2155-9554.1000366

Case Report Open Access

Poromatosis Following Ewing Sarcoma

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Received date: June 21, 2016; **Accepted date:** August 09, 2016; **Published date:** August 11, 2016

Abstract

Eccrine poromas are rare, benign adnexal tumors derived from the intraepidermal portion of sweat ducts. Historically they were thought to arise from eccrine ducts although today it is thought that they may also have an apocrine origin. They usually appear as solitary, slow-growing, skin-colored papules on acral surfaces. Here we present the unusual situation of a patient with multiple poromas who was previously treated with chemotherapy and radiation for Ewing sarcoma. This report adds to the increasing evidence that connect multiple poromas to some treatments of malignant conditions.

Keywords: Ewing sarcoma; Poroma; Poromatosis

Introduction

A poroma is a benign adnexal neoplasm composed of epithelial cells that show tubular differentiation (usually distal ductal), whereas a porocarcinoma is a malignant poroma which can cause visceral metastasis [1]. Historically poromas were considered as glandular adnexal neoplasms of eccrine lineage, and that is why it was referred to as eccrine poroma. However, nowadays it has been proved that this description was not accurate and poromas can be of apocrine lineage too [2]. Multiple poromas or eccrine poromatosis is an uncommon phenomenon and more than half of the reported cases had a history of immunosuppression from either radiation or chemotherapy [3]. Here we present a 51-year-old gentleman with eccrine poromatosis, who was also treated with chemotherapy and radiation before the onset of the lesions.

Case Report

A 51-years-old Saudi male with a history of Ewing sarcoma of the chest, which was diagnosed in April 2012 in the university hospital. He presented initially with 3 months history of painless gradually enlarging right chest mass. No other complaints and systemic review was unremarkable. Blood studies were within normal parameters. A chest X-ray showed opacity in the right side of the chest. Computed tomography was ordered for further delineation of the opacity and revealed a large well defined lobulated heterogeneous solid enhancing mass with central low density area measuring 5.1 x 6.6 cm lying subcutaneously in the right anterior chest wall and reaching the lower level of sternum. There was no evidence of mediastina or other adenopathies. A diagnosis of primitive neuroectodermal tumor (PNET) of the chest wall was made through a CT-guided biopsy of the mass. An isotope bone scan confirmed that no other skeletal site was involved. The patient underwent surgery on May 3, 2012, in form of wide local excision of the right chest wall mass and rhomboid flap. Histopathology of the mass showed no evidence of the neoplasm extension to the right chest wall muscle. The tumor was classified as stage 1b. After the surgery, fractionated radiotherapy of 45 Gy was decided, but the patient was not able to continue for social reasons after receiving two sessions. Chemotherapy has been offered and he found it more convenient for his situation. He received a regimen consisting of Etoposide, Ifosfamide and Mesna starting on June 5, 2012. He was given 4 cycles of chemotherapy with the last one on October 19, 2012. A repeat CT scan did not show any recurrences. To date, the patient remained disease-free after 4 years of follow-up in the oncology clinic. The patient was seen at the dermatology clinic in July 2015 because of eruptive multiple papules and nodules on the chest and abdomen. These lesions had developed over the past year and are asymptomatic. On examination the patient looks fine, although he has a long scar from his previous surgery over the right chest and flank. He has multiple erythematous to brown, mildly firm papules and nodules over the anterior trunk ranging in size between 0.3 to 1 cm (Figures 1a and 1b) involving the chest and abdomen. The back, extremities and other body sites are free of such lesions. A review of systems was unremarkable.

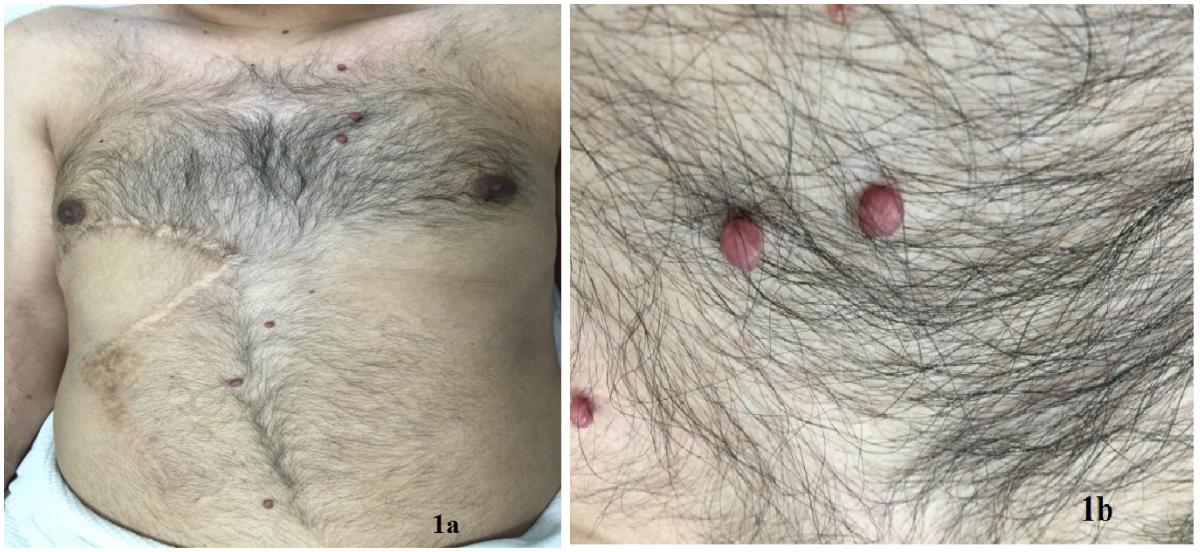

Figure 1: (a) Multiple erythematous to slightly brown firm papules and nodules over the anterior trunk; (b) closer view of the poromas on the chest.

A biopsy was taken and histological analysis revealed well-circumscribed tumors composed of nests and cords of proliferating basaloid cells. The cells were uniform and smaller than the epidermal cells with which they were in contact. The stroma was vascular and ducts were seen in the tumor nests (Figures 2a-2c). Based on those features, a diagnosis of multiple poromas was made. All the lesions were excised and sent for pathology, and sections from all lesions confirmed the same diagnosis. There was no distinctive histopathological feature differentiating these poromas in our patient from the typical de novo lesions. Patient was seen after 1 year of lesions removal with no recurrences or new lesions.

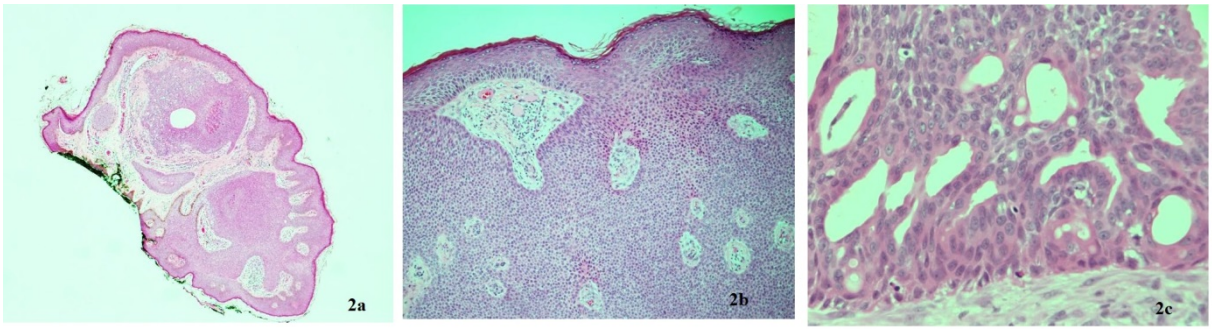

Figure 2: (a) Biopsy of one of the nodules shows circumscribed tumors composed of nests of basloid cells extending downward from the epidermis. Hematoxylin and eosin x40; (b) Medium magnification of the tumor shows uniform small cells and vascular stroma. Hematoxylin & eosin x100; (c) Ducts present in the tumor. Hematoxylin & eosin x400.

Discussion

Eccrine poroma (EP) is a benign, mostly asymptomatic, slow growing or stable neoplasm appearing as nodular lesions, occasionally pigmented, which occurs typically on the palms and soles [4]. Its pathogenesis remains unclear. The earlier literature explains that by the high concentration of eccrine glands on the acral surfaces, especially the sole of the foot. However, newer reports demonstrate that it can form elsewhere on the body, including areas without a high density of eccrine ducts and in hair-bearing areas [5,6]. It arises within the epidermis and it is superficial, with sharply demarcated borders, originating from the uppermost portion of the intra-epidermal eccrine duct and the acrosyringium [7]. As the lesion develops, the tumor starts to develop in the dermis layer. Poromas usually present as solitary papules, plaques or nodules and it can appear on any cutaneous surface, where they are less readily specifically identified. Another common site is the scalp, with or without an associated nevus sebaceous. EPs have been reported in all age groups, including children [5,8]. Poromas commonly display highly vascularized stroma, and a clinical pattern suggestive of pyogenic granuloma is common, especially in poromas that involve acral skin. Due to their benign nature treatment can be optional, although some clinicians emphasize the importance of excision to avoid possible future malignancy [2]. Superficial lesions may be treated by shave or electrosurgical

destruction. Superficial or deeper lesions may also be treated by simple excision.

In histologic examination, poroma consists of a circumscribed proliferation of small, compact, pale cuboidal epithelial cells with monomorphous nuclei and scant eosinophilic cytoplasm. The pale-staining aspect of these cells distinguishes them from the dark-staining cells of basal cell carcinoma (BCC). Poromas can be wholly intra-epidermal, a pattern known historically as hidroacanthoma simplex; can occur in nests and cords in the dermis with broad continuity with the epidermis and are then known as juxtaepidermal poroma, or they can develop wholly within the dermis and are known as intradermal poroma, a pattern known historically as a dermal duct tumor.

Poromas may occasionally display tubular foci in which a central cuticle is lacking. If such tubular foci are lined by columnar cells with holocrine secretion at the luminal border, a presumptive diagnosis of apocrine poroma can be offered [4]. Many other evidences now suggest that some of these lesions may actually be derived from the apocrine gland rather than the eccrine gland [2].

Although poromas can usually be deemed benign, based on circumscription and benign cytologic features, these lesions not uncommonly show small foci of necrosis en masse and a highly vascularized (granulation tissue–like) stroma, the combination of which can create a concern for malignancy, especially in inexperienced observers.

In rare reports, multiple poromas can develop, either in an acral location or in a widespread distribution, a clinical pattern referred to as poromatosis. One old report in 1970 described an acral type of poromatosis but did not mention if that patient has any underling medical problems that could be related [9] Wilkinson et al. described a patient with hidrotic ectodermal dysplasia presented with diffuse poromatosis involving the trunk and extremities, including palms and soles [10]. Four cases of eccrine poromatosis have been reported in association with radiotherapy or chemotherapy in patients diagnosed with different kinds of malignancies including osteomyelitis, mycosis fungoides, acute lymphocytic leukemia and non-Hodgkin's lymphoma [5,11-13].

In another recent paper, three patients were reported with non-Hodgkin's lymphoma and one with malignant fibrous histiocytosis, who were all treated with chemotherapy and presented after various periods of time with multiple poromas [14]. They stated that chemotherapy is a well-documented cause of various cutaneous disorders, such as neutrophilic eccrine hidradenitis (NEH) and syringosquamous metaplasia, probably due to the concentration of chemotherapeutic agents or their metabolites in the sweat glands and ducts as described in the literature before. They noticed long latency periods between the end of chemotherapy and the diagnosis of eccrine poroma and suggested the possibility that the development of eccrine poroma is associated with remodeling of the sweat apparatus or the regeneration of damaged skin appendages.

In 2013, as reported in the American journal of Dermatopathology, a 72-year-old woman underwent radical excision of an adenocarcinoma of the right nasolacrimal duct and eruptive poromas occurred soon after radiation therapy. The patient was not treated with chemotherapy. They concluded that the occurrence of multiple poromas may be related to immunosuppression [15].

In our report, the presentation started almost 3 years after the chemotherapy and radiotherapy but still we think it is related to them, as longer latency periods were the rule in most of the previous reports. It was hard to know which of them could be the most responsible as both modalities were used in our patient, and even harder to know which single chemotherapeutic agent most likely related to development of poromas. Patient withdrawal from radiotherapy after only two sessions could raises more suspicion toward the chemotherapy. Although the lesions developed mostly nearby but outside the field of radiotherapy, literature has described eruptive poromas after radiotherapy both at the irradiated field and in non-treated areas [12,15]. After reviewing the cases reported association with chemotherapy, ifosfamide and its parent analogue cyclophosphamide have been mentioned in 6 cases including our case, while doxorubicin and its derivative epirubicin in 5 cases. Although it is difficult to assert their causality, cyclophosphamide and doxorubicin would be the most suspected agents associated with poromas. Vincristine has been mentioned in 4 cases [5,14]. Another case reported association with total body irradiation in patient with leukemia also treated with chemotherapy, but the chemothraputic agents were not mentioned in this article [13].

The prognosis of poromas is favorable and even poromatosis is not known to be associated with other anomalies. However, given the rarity of these neoplasms, there remains a paucity of information on atypical presentations, recurrence and rates of malignant transformation. Eccrine porocarcinoma may arise de novo or may develop from a pre-existing poroma. The variation in the reported rates of malignant transformation from benign poromas is very evident. It has been reported to be as high as 100%, while others reported only 18% of poromas become malignant. The progression to malignancy takes a mean of 8.5 years [16]. If the lesion recurs after excision or pre-existing poroma presents with spontaneous ulceration, bleeding, pain, sudden itching or accelerated growth, eccrine porocarcinoma should be suspected. It tends to appear more exophytic and ulcerative than a benign poroma. Our patient had no recurrences or new lesions after 1 year of follow up and he will be under annual reassessment.

In conclusion, our report will add to the increasing evidence in the literature that connect multiple poromas to some sort of treatments, especially for malignant conditions. Finally, these reports suggest that chemotherapeutic agents and radiotherapy could be responsible for the occurrence of the multiple eccrine poromas.

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