

Fibroepithelioma of Pinkus Masquerading as a Widely Distributed Papulosquamous Disease

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Abstract

Fibroepithelioma of Pinkus (FeP) is generally considered a rare tumor that usually presents as a solitary lesion. Multiple tumors are extremely exceptional and usually associated with certain risk factors. We are presenting a peculiar case of FeP presenting with multiple scaly erythematous papules and plaques masquerading as a papulosquamous disease. This patient had no risk factors that could predispose him to develop multiple skin tumors. Up to our knowledge, the case we are reporting is the fourth case in literature of multiple FeP without a history of antecedent radiation and occurring independently of genetics diseases predisposing to skin malignancy.

Keywords: Fibroepithelioma of Pinkus, multiple lesions, no risk factors

INTRODUCTION

The case we are presenting is interesting because up to our knowledge and based on the extensive searched done, it presents the fourth case of multiple FeP without any known risk factors.

CASE REPORT

We are reporting a 62-year-old male presented with asymptomatic multiple skin lesions, occurring bilaterally on lower extremities and gradually increasing in size over a period of 4 years. The clinical examination revealed a sharply demarcated erythematous scaly papules and plaques of a variable sizes ranging from 5 mm to 10 mm in diameter. There was an ulcerated red-brown well-defined papule (5 mm × 5 mm) on the face [Figure 1]. The differential diagnosis was psoriasis, pityriasis lichenoides chronica, or lichen planus. The dermoscopic examination showed fine arborizing vessels, white streaks, and an irregularly distributed brown structureless area of pigmentation [Figure 2]. The histologic examination of a 4 mm punch biopsy showed a thin anastomosing strands of basaloid cells projecting from the epidermis in a fenestrated pattern. The cords of a peripheral palisading basaloid cells are embedded in abundant fibrous stroma. Based on the microscopic features, the diagnosis of FeP was made [Figure 3]. However, due to the rarity of FeP occurring as a multiple lesions, conformational biopsies were taken from the leg and

the face. The result was consistent with the initial histologic diagnosis, however, the facial lesion turned out to be basal cell carcinoma (BCC). Detailed medical and family histories were unremarkable. There was no history of exposure to radiation or ingestion of potential carcinogenic material. Full skin and systemic examination were normal. Imaging studies, namely X-rays of the skull, chest, and jaw, as well as an ultrasound of abdomen and pelvis, were normal. All lesions were excised surgically followed by primary closure. Follow-up visits were arranged monthly for the first 3 months then every 6 months, during which the patient was clinical free.

DISCUSSION

FeP is considered a relatively rare tumor.^[1] According to BCCs series, the frequency of FeP ranges from 0.2% to 1.4%.^[1] However, the true incidence of FeP may be underreported, as FeP is frequently mistaken for a variety of benign lesions which may not be treated or biopsied.^[1,2] Most FePs appear in 4th-6th decade,^[1,2] few cases were reported in pediatric population.^[1] FeP has slightly higher prevalence among females.^[1]

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How to cite this article: Alramthan A, Elbohy M, Shaban M, Alenezi M. Fibroepithelioma of pinkus masquerading as a widely distributed papulosquamous disease. *J Dermatol Dermatol Surg* 2019;23:49-51.

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DOI:
10.4103/jdds.jdds_52_18



Figure 1: A sharply demarcated multiple erythematous scaly papules and plaques of a variable sizes ranging from 5 mm to 10 mm in diameter



Figure 2: Fine arborizing vessels and dotted vessels, white streaks, and an irregularly distributed gray-brown structureless area of pigmentation

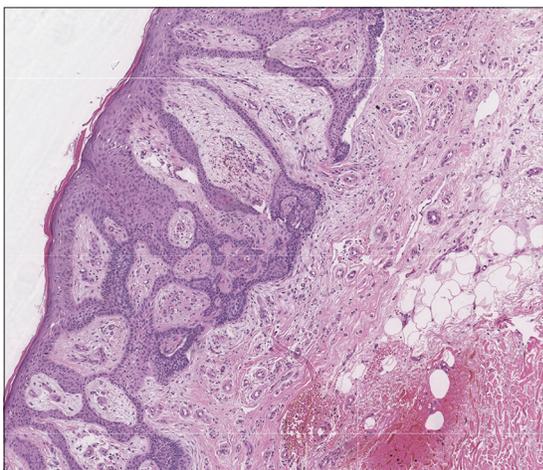


Figure 3: Cords of basaloid cells are embedded in abundant fibrous stroma. The cells at the edge of cords show peripheral palisading

FeP usually presents as a asymptomatic, solitary, firm, skin-colored or slightly brown-gray, fleshy, sessile, or pedunculated papule with a broad base. Infrequently, it may

present as ulcerated nodule.^[1,2] Presentation as multiple lesions is rare.^[1,2] FeP occurs mainly on the trunk with predilection for the lumbosacral area, but it may arise in atypical locations such as extremities, axillae, groin, head, genitalia, and planter foot.^[1,2] In contrast to BCC, FeP shows no predilection for sun-exposed sites.^[1] Yet, it tends to develop more often in patients with a history of BCC.^[1] FeP may mimic various benign conditions, for example, acrochordon, pedunculate fibroma, neurofibroma, or seborrheic keratosis (SK). It may also be mistaken for amelanotic melanoma.^[1]

FeP is characterized histologically by numerous, elongated, 2–3 cell thick anastomosing strands of basaloid cells projecting downward from the epidermis into the dermis within a fibrotic stroma, giving the tumor a honeycomb appearance.^[1,2] Histological differential diagnosis includes reticulated SK, tumor of follicular infundibulum, eccrine syringofibroadenoma, and mammary intracanalicular fibroadenoma.^[1] It occurs not only in a pure form but also in combination with another form of BCC.^[1]

Dermoscopy of FeP reveals polymorphous vascular pattern consisting of fine short arborizing vessels either alone or in association with dotted vessels, and shiny white streaks, known as crystalline structures, that are only visible on polarized dermoscopy.^[2] Gray-brown irregularly distributed structureless pigmentation associated with a variable number of gray-blue dot characterizes the pigmented FePs.^[2] Additional features may include milia-like cysts and ulceration.^[2] A recently reported feature that may be more specific for FeP is the white network. It correlates with a regular anastomosing network of epithelial cells strands of emanating from the undersurface of epidermis.^[3]

FeP has an indolent course. Aggressive behavior, local destruction, or metastasis is extremely rare.^[1,2,4] Complete surgical excision of tumor is curative.^[1,2] Other treatment modalities include electrodesiccation and curettage, cryosurgery, or Mohs surgery.^[1,2] Topical imiquimod 5% is ineffective in treating FeP.^[2]

FeP has been documented to occur in association with previous exposure to radiation in various publications.^[4-6] The degree to which radiation contributes to the development and evolution of FeP is not clear.^[6] Predisposing genetic factors should be suspected in patients with multiple, widespread, and early-onset lesions. Some syndromes are associated with multiple BCCs, including FePs, namely Gorlin, Bazex, and Rombos syndromes.^[7]

We reviewed the published literature in an attempt to assess the incidence of multiple FeP. The term “Pinkus tumor,” “fibroepithelioma BCC,” and “FeP” have been searched in PubMed without restriction up to March 2018. Four hundred and ninety-nine published articles associated with FeP were found. A total of six articles only reported FeP presenting as multiple lesions.^[4-10] Majority of these cases were associated with exposure to radiation.^[4-6] Only three articles reported multiple FeP occurred in isolation of any inherited genetic

abnormality and radiation exposure.^[8-10] Sehli Attafi *et al.* described a 55-year-old male with multiple lesions that clinically resembled SK, but histological confirmed to be FeP.^[8] Barr *et al.* reported an 80-year-old male with four FeP. He mentioned that multiple FeP accounted for 36% of the total reported cases at that time. However, there was no specification to whether this percentage included cases with antecedent radiation or genetic abnormalities.^[9] Rodriguez and Festa Neto reported multiple FeP occurring in a nevoid distribution associated with SK.^[10]

The case we are presenting is interesting because up to our knowledge and based on the extensive searched done, it presents the fourth case of multiple FeP without a history of antecedent radiation and occurring independently of genetic diseases predisposing to skin malignancy. In addition, it has atypical clinical presentation and uncommon distribution.

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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