BRIEF ARTICLES

Fibroelastolytic Papulosis in a Middle-Age Female: Presentation and Review of Treatment

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ABSTRACT

Fibroelastolytic papulosis (FEP) is a rare, benign, acquired cutaneous disease with a histopathology that shows variable fibrosis and elastolysis of the papillary dermis. FEP clinically presents as white-ivory to yellow papules and plaques commonly occurring on the neck. There are no widely accepted treatment guidelines and several management options will be discussed. We present a 62-year-old female with an isolated ivory, cobblestoned plaque with open comedones on the left shoulder since childhood. The histopathology confirmed the diagnosis of FEP. The patient had been previously treated with topical clindamycin, salicylic acid, tretinoin, and tazarotene without success. This case demonstrates the importance of recognizing FEP, as clinical presentations can vary. FEP can be distressing to patients, and it is important to explore additional treatment options. Treatment options including topical retinoids and ablative lasers have been reported, but with limited and inconsistent success. However, due to the rarity of the disease there is currently no standard of care for the treatment of FEP and additional successful treatment options are needed.

INTRODUCTION

Fibroelastolytic papulosis (FEP) is a rare acquired, non-systemic, skin disease that presents as white-ivory to yellow papules and plaques commonly occurring on the neck (1). In 1989 Shimizu et al. reported an isolated asymptomatic white fibrous plaque with histopathological changes of fibrosis in the papillary and upper reticular dermis which was coined as white fibrous papulosis of the neck (WFPN) (2). In 1992 Rongioletti and Rebora reported a similar skin lesion identifying it as pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE), which, unlike PXE, does not have systemic associations (3). Balus et al. first proposed the name “fibroelastolytic papulosis of the neck" (FEPN) in 1997 to encompass both WFPN and PXE-PDE, since they believed these skin disorders were a continuum of a single entity (4,5). Despite the name, FEPN can occur in other locations besides the neck, thus Jagdeo et al. recommended changing the name to solely “fibroelastolytic papulosis (FEP)” in 2004 (6).

Clinically, FEP presents as multiple (20-100), 4-6 mm firm papules often coalescing into a

cobblestone plaque. They range in color from flesh colored to white-ivory to yellow. The papules and plaques are generally asymptomatic, but inflamed folliculitis-like areas and pruritus can occur. FEP is found on the posterior and lateral aspects of the neck with extension to surrounding areas. The histopathology of FEP presents as loss or alteration of elastic fibers and fibrosis of the papillary and upper reticular dermis(7). PXE and PXE-PDE will reveal calcified elastic fibers(3).

The pathogenesis of FEP has yet to be elucidated, but it is commonly theorized that it is related to an intrinsic photo-aging process. FEP most often occurs in patients over the age of 40 with a predilection for females of European ancestry (7). The lesions of FEP are benign, but can often progress and be cosmetically distressing to patients. There are no standard FEP treatments established at this time due to its rarity. We report an interesting case of FEP in a 62-year-old Caucasian female and explore treatment options.

CASE REPORT

A 62-year-old female patient with a past medical history significant for hypothyroidism and hyperparathyroidism presented with a large lesion on her left shoulder since childhood. The patient described it as generally asymptomatic but reported intermittent pimple-like lesions within the plaque that developed later in life.

Physical exam revealed multiple 2-4 mm flesh-colored to ivory papules coalescing into a large cobblestone, well-defined plaque with inflammatory papules and open comedones on the left shoulder extending on to the left upper back and proximal left upper extremity (Figure 1).

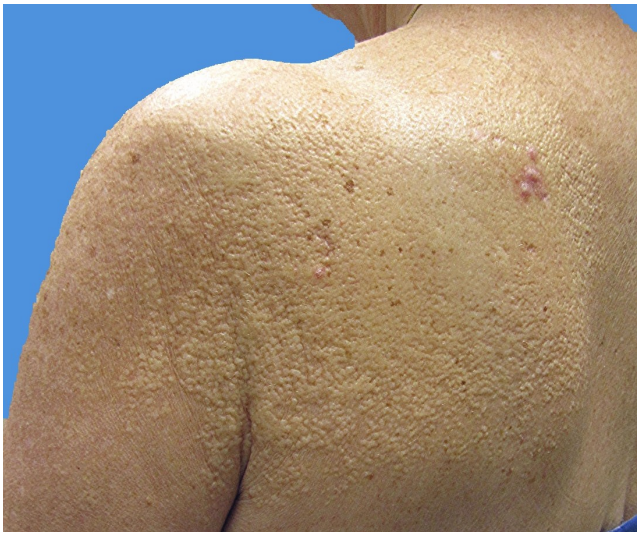


Figure 1: Clinical image demonstrating multiple 2-4mm fleshed colored to ivory papules coalescing into a large cobblestone-pattern plaque with dilated comedonal openings on the left shoulder and proximal left arm.

The histopathology revealed intradermal proliferation of variably sized tubular structures lined by a multi-cell layered epithelium and filled with amorphous basophilic material. Distinct small areas of superficial dermal collagen thickening with some of the collagen bundles becoming hyalinized, thickened and amorphous were present (Figures 2, 3). Focal patchy inflammation was noted within portions of the dermis surrounding the hyalinized nodules. Considering the clinical presentation and histopathological correlation, the diagnosis of FEP was made.

Due to the acneiform nature of the lesions within the plaque the following treatments were attempted; topical clindamycin 1% lotion, salicylic acid wash, benzoyl peroxide 5% wash, tretinoin 0.5% cream, and tazarotene 0.1% without considerable improvement. Patient refused all additional treatment including fractionated carbon dioxide laser treatments.

Figure 2: Hematoxylin and Eosin (4x) slide from a punch biopsy of left mid superior scapula demonstrating superficial dermal collagen thickening, some of which are sharply defined, with some of the collagen bundles becoming hyalinized, thickened and amorphous. Some focal patchy inflammation is noted within portions of the dermis surrounding the hyalinized nodules as well.

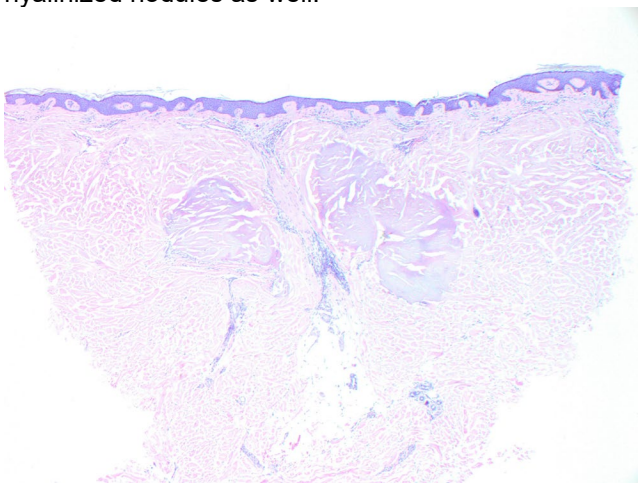
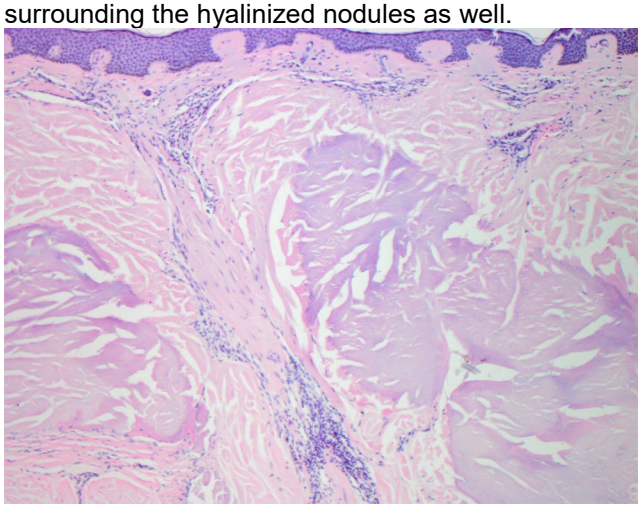


Figure 3: Hematoxylin and Eosin (20x) slide from a punch biopsy of left mid superior scapula demonstrating hyalinized, thickened and amorphous superficial dermal collagen. Some focal patchy inflammation is noted within portions of the dermis surrounding the hyalinized nodules as well.



DISCUSSION

FEP is a rare acquired skin disorder that shows elastolysis of the papillary dermis. FEP encompasses WFPN and PXE-PDE due to considerable overlap in clinical presentations and histopathology. The pathogenesis of FEP is not well understood, but may be secondary to intrinsic and photo-aging processes (8). FEP is a non-life threatening skin disease, nevertheless it can be distressing for many patients due to cosmesis. In the case reported the patient experienced discomfort due to inflammatory lesions. There are currently no standard treatment guidelines established for FEP.

Considering the benign nature of FEP, treatment is not necessary. However, many patients are cosmetically concerned with FEP and seek treatment. Tretinoin may be a viable non-invasive, well-tolerated option for patients with FEP, but results are inconsistent and often not curative. One case report documented the use of topical tretinoin 0.05% cream twice a day with significant improvement after five weeks (8). Similarly, another case of FEP treated with topical tretinoin 0.1% gel improved after three months and revealed complete resolution in one year (9). In contrast, a case reported by Lueangarum, et al., did not improve with tretinoin in their patient (10).The patient we presented also did not show improvement with tretinoin. The exact mechanism by which tretinoin acts on FEP is unknown (8).

Moreover, two case reports, one by Chong et al., and the other by Ho et al., documented successful treatment of FEP with fractionated carbon dioxide (CO2) laser (SmartXide DEKA DOT® Laser)(1) (11), with improvement after three successive laser treatments and complete resolution within a year. Another case report highlighted the successful treatment of FEP utilizing a non-ablative fractional photo thermolysis laser (Fractionated 1550-Erbium Glass laser) (10). Based on these findings, it appears that laser treatment may be a viable treatment option for FEP. Surgical excision can also be

considered in a small well-defined lesion of FEP (10).

CONCLUSION

Fibroelastolytic papulosis (FEP) is a rare benign skin disorder that can be distressing to patients, and has no established treatment guidelines to date. Our case demonstrates a unique clinical presentation and the need to explore additional treatment options for FEP. There have been few successful FEP treatments documented but ablative lasers appear promising. Despite inconsistent results with topical retinoids, this modality could also still be considered as a conservative first line approach to therapy. No single treatment has been thoroughly studied for its efficacy in FEP, therefore treating each case on an individual basis with patient consideration is recommended.

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