Multiple Eruptive Myxoid Dermatofibroma in a Male with Chronic Hepatitis B Infection

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ABSTRACT

We report the first published case of multiple eruptive myxoid dermatofibroma (MEMDF) in a male with chronic hepatitis B infection presenting with eruptive lesions showing marked deposits of dermal mucin. Alcian blue and immunohistochemistry confirmed the diagnosis of myxoid dermatofibroma. Further work-up showed asymptomatic chronic hepatitis B infection without cirrhosis.

This case highlights an extremely rare histologic variant and the importance of screening for altered immunity in patients with eruptive dermatofibromas.

Key Words: dermatofibroma, eruptive dermatofibroma, myxoid dermatofibroma, multiple eruptive myxoid dermatofibroma

Introduction

Dermatofibromas are among the most common clinical and histopathologic diagnoses seen in dermatologic practice, presenting as solitary lesions on the extremities with a female predominance.

Rarely, they may occur in multiples over a short period of time, as multiple eruptive dermatofibromas (MEDF). These conditions have been found on a background of altered immunity,[1,2,3] most commonly systemic lupus erythematosus (SLE) and the human immunodeficiency virus (HIV).

Histologically, dermatofibromas are characterized by fibrohistiocytes infiltrating thickened collagen bundles. The finding of marked stromal mucin deposition, in myxoid dermatofibromas, is an extremely rare variant, accounting for 0.4% of all lesions.

To date, only two cases of multiple eruptive myxoid dermatofibromas (MEMDF) have been published in literature[1,4] – one with no underlying disease and one with SLE (Table 1).

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We report another rare case with both the rare clinical presentation of eruptive dermatofibromas and the extremely rare histologic myxoid variant, this time, in a male with chronic hepatitis B.

Case Report

A 55-year-old Filipino male came in for multiple papules and nodules over the right upper back starting one year prior with a solitary erythematous papule over the right upper back gradually becoming firm, nodular, and hyperpigmented, and developing dull pain over a few months. He denied any history of antecedent trauma to the affected area. There was no associated pruritus, fever, weight loss, or malaise.

In the interim, newer papules and nodules were noted on the right back.

The patient was hypertensive with no other known comorbidity. He had no family history of skin tumors or autoimmune disease. He denied a history of alcoholism.

The patient was previously treated for an episode of genital discharge. His first born was recalled to be HBsAg-reactive at birth, while the patient's spouse claims being generally healthy with no recalled laboratory abnormalities during pregnancy.

On examination, we noted 27 circumscribed well to poorly-defined pink-red to tan-brown papules and nodules over the right shoulder and back with extension to the posterior and mid-axillary line (Figure 1). Lesions ranged from 1 mm to 12mm in widest diameter. Some were dome-shaped, polypoid, and flat. No ulceration, bleeding, or telangiectasia was noted. Tenderness was elicited over the more hyperpigmented and larger nodules. Fitzpatrick sign was negative for all lesions. Dermoscopy on lesions of different morphologies showed delicate and atypical pigment networks at the periphery and central hyper- and hypopigmentation.

A 4-mm punch biopsy on the largest and oldest nodule on the back revealed a circumscribed poorly defined lesion within filtrative borders, a spindle cell proliferation of fibroblasts wrapping around thickened collagen bundles, and mild deposition of mucin in the dermis. Epidermal hyperplasia, irregular areas of acanthosis, and basal hyperpigmentation were also noted.

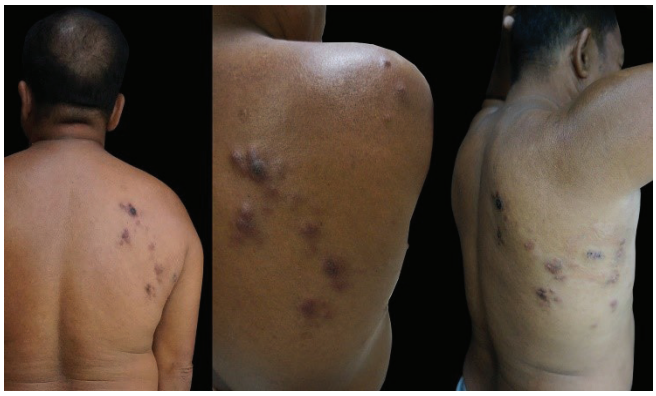

Figure 1. Multiple circumscribed well to poorly defined pink-red to tan-brown papules and nodules over the right shoulder and back with extension to the posterior and mid-axillary line.

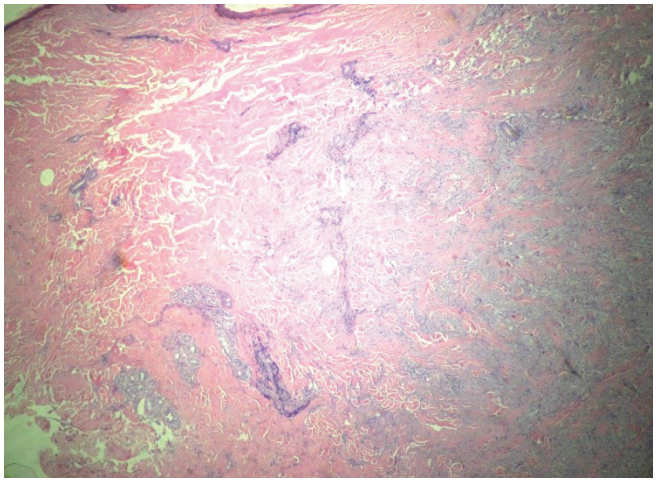

Figure 2. H&E stain showing a well-circumscribed dermal nodule exhibiting epidermal hyperplasia and fibroblasts wrapping around thick collagen bundles (10x).

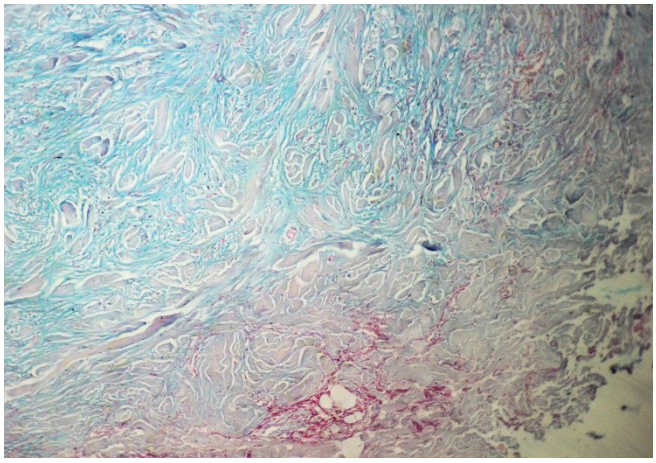

Figure 3. Alcian blue stain confirming marked deposits of mucin in the dermis (10x).

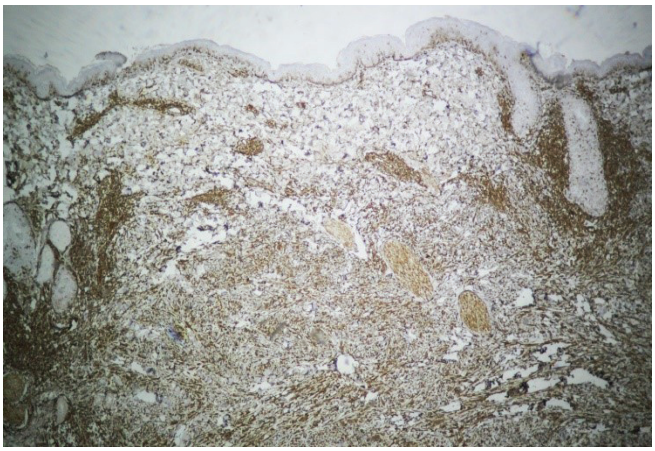

Figure 4..Vimentin, staining structures of mesenchymal origin, is positive (10x).

Figure 5. CD68-positive staining, labeling fibroblasts (10x).

Excision of a papulonodule on the posterior axillary line showed a well-circumscribed dermal nodule exhibiting similar findings to the punch specimen, ie, epidermal hyperplasia, fibroblasts wrapping around thick collagen bundles (Figure 2). Marked deposits of mucin were appreciated within the dermis, confirmed by positive staining for Alcian blue (Figure 3).

Immunohistochemical staining was positive for vimentin and CD68 (Figure 4 [vimentin] and Figure 5 [CD68]); negative results were noted for S-100 protein, and smooth muscle actin (SMA). CD34 revealed regular staining of endothelial cells.

The patient was negative for markers of SLE and HIV. However, he was seen to have chronic hepatitis B infection with no signs of active infection and cirrhosis. Liver function tests were normal. Whole abdominal ultrasound showed hepatomegaly with fatty changes with no evident masses or nodules. He was advised biannual follow-up to monitor hepatic status and lifestyle and diet modification.

Table 1. A review of patients with multiple eruptive myxoid dermatofibromas and associated conditions

Reference	Distribution	Sex/age (years)	Underlying disease	Immunohistochemistry
Antal et al (2007)[1]	Generalized	F/20	None	**Positive:** vimentin **Negative:** CD34, S-100, CD68, SMA
Wu et al (2015)[4]	Arms, legs	F/51	SLE	**Positive:** Factor XIIIa, CD68, CD44, CD163 **Weakly positive:** NK1/C3 **Negative:** S-100, CD34, caldesmon, collagen type IV

Although no treatment is required for asymptomatic dermatofibromas, excision of the few other symptomatic lesions was offered to the patient. Due to uncontrolled blood pressure elevations, the procedure was postponed.

Discussion

The histopathological findings of circumscribed nodules with spindle-shaped cells in a collagenous stroma and infiltrative borders of fibroblasts wrapping around thickened collagen bundles were strongly suggestive of dermatofibroma as the diagnosis. That it was vimentin and CD68-positive confirmed it was a spindle cell tumor. Marked mucin deposits in the dermis were confirmed by Alcian blue, suggesting a myxoid variant.

Other tumors of spindle cell origin (eg, dermatofibrosarcoma protuberans, neurofibromas, fibromyxoids arcoma, and pilarleiomyoma) were ruled out by negative CD34, S-100, and SMA immunoreactivity.

Baraf and Shapiro[5] initially defined multiple dermatofibromas as the presence of at least 15 lesions; however, the relevance of this number is still in question.[1,3] Rather, it has been proposed that multiple eruptive dermatofibromas be defined as the presence of 5 to 8 DFs appearing within a period of 4 months.[1] Nonetheless, the clinical scenario of the patient fits this criteria.

Dynamic changes in some lesions within a short period of time, in contrast to the static state observed in common dermatofibromas, are characteristic of multiple eruptive dermatofibromas (MEDF).[3]

A review of 106 patients with MEDF made by Niiyama[3] in 2002 found that more than half of cases were associated with systemic disease (56%). Among those with systemic diseases, SLE (46%) and HIV (32%) were leading co-morbidities, followed by other immune-mediated diseases, such as myasthenia gravis and pemphigus vulgaris. This predominance of SLE and HIV among patients with MEDF was confirmed by a review by Wu.[4]

Myxoid dermatofibroma is found in 0.4% of all dermatofibromas.[6]

After searching possible cases through electronic search engines and reference lists, we found only two published reports on multiple eruptive lesions with histopathologic findings consistent with myxoid dermatofibroma[1,4] (Table 1). In these reports, both patients were females – one with SLE and one with no underlying disease.

This case is a noteworthy addition to literature because of the unusual clinical and very rare histologic presentation of dermatofibroma. To the best of our knowledge, this is also the first case of a male with multiple eruptive myxoid dermatofibromas, which occurs on a background of chronic hepatitis B without evidence of cirrhosis.

Statement of Authorship

All authors have approved the final version submitted.

Author Disclosure

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