

**Case Presentation**

**Multiple clustered and focally atrophic dermatofibromas (DF)**

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**Abstract**

Multiple clustered dermatofibromas describes a confluence of dermatofibromas in one anatomic location. We describe a 32-year-old man who presented for evaluation owing to skin papules and plaques and concerns about malignancy. Repeat histopathological evaluation found no evidence for dermatofibrosarcoma protuberans. Our case presents this relatively rare condition and discusses observation along with potential treatment options.

**Case synopsis**

A 32-year-old man initially presented in 2009 with a reticulated red-brown plaque on his left thigh that had been present and stable for approximately 15-years. However, 4-months prior to presentation it progressively expanded. The patient denied a history of trauma; other than occasional pruritus, the lesion was asymptomatic. A punch biopsy was performed, but the patient was lost to follow-up. The patient recently returned to clinic for re-evaluation. On physical examination, there was an ill-defined 10 x 20 cm region on the left anterior thigh composed of multiple red-brown plaques that varied in size and shape with intervening areas of normal appearing skin (Figure 1). The epidermis overlying some of the plaques had a wrinkled appearance and some plaques were focally depressed (Figure 2). Reduction of terminal hairs was also noted in this region. An incisional biopsy was performed (Figure 3 and 4).

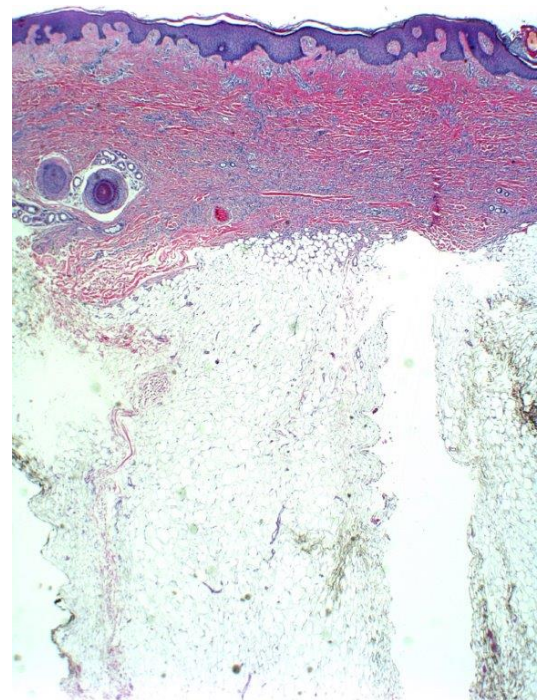
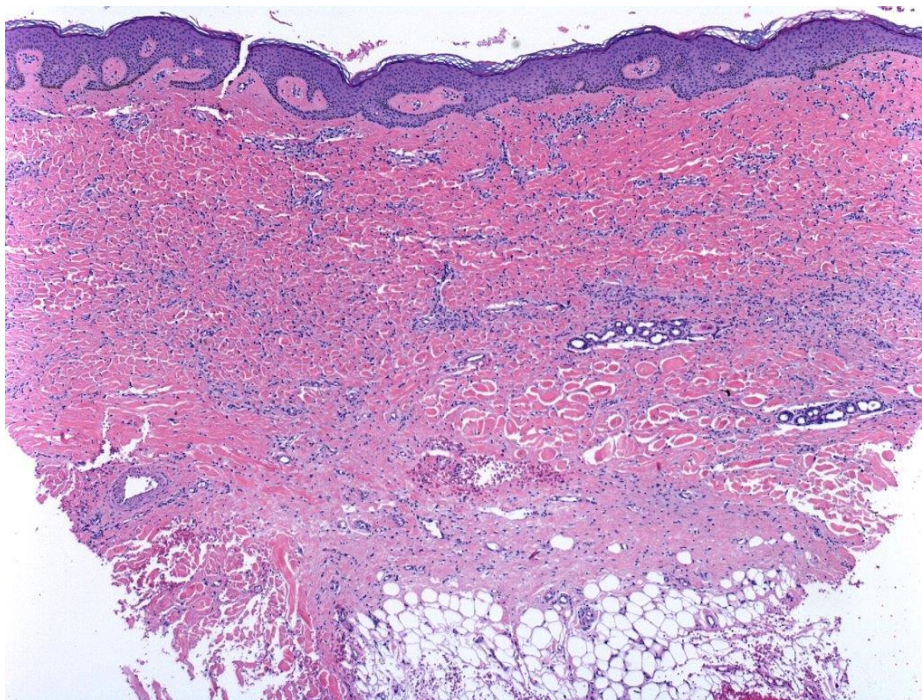


**Figure 1.**



**Figure 2.**

Histological evaluation revealed a dermal spindle cell proliferation associated with peripheral collagen trapping (Figure3). Focal areas showed marked reduction in the dermal thickness (Figure 4). The overlying epidermis was acanthotic with increased basal layer pigmentation. The cells of interest diffusely labeled with Factors XIIIa and failed to stain with CD34. Prior biopsies were reviewed and showed similar histological findings. Although the lesion was consistent with multiple clustered and focally atrophic dermatofibromas, owing to its unusual appearance a MRI was performed, which showed no evidence suggestive of a dermatofibrosarcoma protuberans (DFSP). No treatment was pursued other than clinical follow-up.



**Figures 3 and 4.**

## **Discussion**

Dermatofibromas are common benign fibrohistiocytic proliferations. Classically they present as small firm nodules slightly tethered to the underlying tissue and they most frequently arise on the extremities of young adults. The lesions are more often solitary. However, 2-5 lesions are present in approximately 10% of patients [1]. Multiple DF, arbitrarily defined as greater than 15



lesions, can either arise abruptly or develop slowly and tend to be diffusely distributed [2]. These lesions primarily occur in patients with altered immunity, such as immunocompromised patients with SLE, HIV, AML, and taking immunosuppressive agents [2,3]. Multiple clustered dermatofibromas, also referred to as agminated fibrohistiocytomas, is a term used to describe a confluence of DFs restricted to an anatomic location. There have been approximately 25 cases reported in the literature [1-9]. In addition, several other reports describe similar lesions, but use the term plaque-like DF or giant DF [5]. With the exception of one case that occurred in a renal transplant patient, all cases occurred in healthy individuals, with the majority occurring on the lower extremities or lower trunk [2,5]. Typically, lesions occur between the first and third decades of life, but 3 congenital cases have also been reported [2,5,6,7]. Atrophic variants account for approximately 2% of all DFs. They present as depressed plaques on the trunk of middle-aged women. Histologically they resemble classic DFs, but additionally show 50% or greater reduction in the dermis [10].

The histological differential diagnosis includes DFSP, dermatomyofibroma, leiomyoma, and, in pediatric cases, plaque-like myofibroblastic tumor. Immunohistochemical stains can assist in differentiating these lesions. Most dermatofibromas stain diffusely with Factor XIIIa and stromelysin-3, whereas DFSP tend to stain diffusely with CD34 [1,2]. Plaque-like myofibroblastic tumor histologically resembles a DF and labels with Factor XIIIa, but also diffusely labels with SMA [8].

To date, although slow expansion of lesions has been noted, no malignant transformation has been reported with durations of follow-up as long as 20 years [9]. All cases of congenital MCDF were stable and asymptomatic until the patient reached puberty, at which time the lesions would tend to grow and develop new satellite papules [6]. This is the first case report of multiple clustered dermatofibromas that also showed focal dermal atrophy.

The etiology and agminated appearance of multiple DF's remains unknown. Even though cryotherapy, intralesional steroids, PUVA, and surgery have been attempted for cosmetic reasons, the results are variable. Therefore, clinical follow-up is the more prudent course [3,5,7].

## Conclusion

Multiple clustered dermatofibromas is a rarely encountered clinical condition and we present a case in which some of the agminated DFs were atrophic. Although changes may occur, there are no reports of malignant degeneration and observation may be the most prudent course of management.

## References

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