CASE REPORT

Atypical Presentation of Dermatofibroma: A Rare Case

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ABSTRACT

Background: Dermatofibroma, also termed benign fibrous histiocytoma, is a common benign fibrohistiocytic tumor that typically presents as a firm reddish-brown nodule on the extremities. Atypical variants particularly atrophic and giant plaque-like forms occurring on the face are exceedingly rare and may be mistaken for inflammatory or malignant dermatoses. We report a 47-year-old woman who presented with a four-month history of an 8 × 4 cm hyperpigmented, indurated, atrophic plaque on the right side of her face, without antecedent trauma. Clinical differentials included morphea, lupus panniculitis, lichen sclerosus et atrophicus, and subcutaneous sarcoidosis. Histopathology revealed dermal spindle cells in a storiform pattern with collagen entrapment, and immunohistochemistry was positive for CD68 and ASMA and negative for CD34 and S100, confirming dermatofibroma. The lesion was completely excised with cervicofacial flap reconstruction. Recognition of such atypical facial variants is essential to guide accurate histopathological diagnosis.

Keywords: Atrophic plaque, atypical presentation, benign fibrous histiocytoma, CD68, dermatofibroma, facial dermatofibroma, giant dermatofibroma, histopathology, immunohistochemistry

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INTRODUCTION

Benign fibrous histiocytoma, another name for dermatofibroma, is a common benign skin tumor[1]. It often manifests as one or more normochromic reddish-brown or dark brown solid nodules (less than 2 cm) that primarily appear on middle-aged people's extremities, with a minor female preponderance. The dermatofibroma causes the distinctive "dimple sign" when two fingers are pressed side to side because the covering epidermis is tethered to the underlying lesion[2,3]. Facial involvement and giant plaque-like dermatofibroma, defined by its size of more than 5 cm, are uncommon and typically represent more aggressive variants that are difficult to treat and often lead to misdiagnosis. Only a few published cases describe the atypical presentations of dermatofibroma[4–6].

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The rationale for reporting this case lies in the diagnostic challenge posed by an atrophic, plaque-like dermatofibroma occurring on the face, a site at which such variants are seldom encountered and at which the morphology closely mimics inflammatory and connective-tissue disorders, increasing the risk of misdiagnosis and inappropriate treatment. The objective of this case report is to describe the clinical, histopathological, and immunohistochemical features of this rare atypical facial dermatofibroma, to highlight key differential diagnoses, and to provide a concise review of the literature that may aid clinicians in the early recognition and accurate diagnosis of similar lesions.

CASE REPORT

A 47 years old woman with no known co- morbidities presented to us with the complaint of a hyper-pigmented depressed lesion on the right side of her face for four months. She had initially developed asymptomatic skin-colored nodular lesions, subsequent to which she noticed gradual stiffening and hardening of that area, along with a slight depression and hyper-pigmentation. Apart from the progressive enlargement of the lesion, no other associated symptoms were noted. There had not even been any history of trauma on that site. No significant past medical, surgical, drug or family history was noted. Upon examination, a single erythematous

Figure 1: Lesion on Face

Figure 2: Lesion on Face

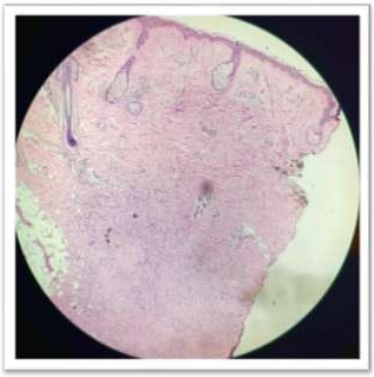

Figure 3

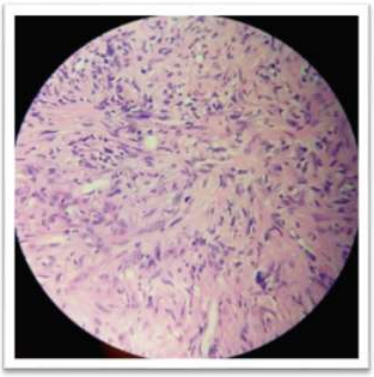

Figure 4

Figure 5: Immunohistochemistry with Positive Asma Stain Which is Made up of Fascicles of Plump, Spindle-shaped Cells with Oval to Irregular Nuclei

Figure 6: Three Weeks After Procedure

to hyper-pigmented indurated and atrophic plaque measuring 8x4 cm with multiple nodules on palpation, appeared laterally on right side of the face just above the lower jaw (Figure 1,2). The rest of the cutaneous and systemic examination was unremarkable.

The initial differentials include morphea, lupus panniculitis, extragenital lichen sclerosus et atrophicus, and subcutaneous sarcoidosis. An incisional skin biopsy was performed. Histopathology showed atrophic epidermis. The reticular dermis shows an ill-defined lesion, which is made up of fascicles of plump, spindle-shaped cells with oval to irregular nuclei and inconspicuous nucleoli.

Intersecting fascicles are arranged in a storiform pattern. Collagen trapping was also evident (Figure 3,4). Immunohistochemical stain CD68 and ASMA were positive, while CD4, CD34, and S100 were negative (Figure 5).

A final diagnosis of dermatofibroma was established based on clinical presentation and characteristic histopathological findings. After counselling the patient about her condition, she was referred to the Surgical Unit for complete excision of the lesion. Finally, a complete excision with cervicofacial flap was undertaken. (Figure 6).

DISCUSSION

Benign fibrous histiocytoma is recognized as one of the most prevalent benign skin tumors, with recurrence rates as low as 3–5%[4]. Dermatofibroma often presents asymptomatically and may appear following minor trauma or an insect bite. It predominantly involves the lower extremities[1].

Several clinical variants of dermatofibroma have been reported in the literature, including atrophic, atypical polypoid, giant, multinodular hemosiderotic, subcutaneous fibrous, keloidal, subungual, generalized eruptive, pleomorphic, ulcerated, erosive, multiple palmoplantar, and multiple clustered varieties.[7] The giant dermatofibroma was initially identified by Danckaert and Karassik. It presents as larger than 5 cm in diameter and appears as an exophytic skin lesion, most commonly affecting the lower limbs, followed by the back[3].

Histopathological analysis reveals that both the classical and giant types of dermatofibroma have several variants, including fibrous/fibrocellular, xanthomatized, aneurysmal, hemosiderotic, epithelioid, cellular, atrophic, lipidized, clear cell, palisading, and keloidal forms[8]. Review of the literature suggests that plaque-like dermatofibromas share a similar histopathological profile with the common fibrous/fibrocellular variant of typical dermatofibroma a poorly defined, non-encapsulated dermal lesion characterized by interwoven bundles of spindle-shaped fibroblasts and macrophages, often arranged in a storiform pattern within a loose collagenous stroma[2,3].

Immunohistochemical staining (IHC) with CD68 highlights the presence of histiocytic cells, while negativity for CD34 helps exclude dermatofibrosarcoma protuberans, which is CD34-positive in approximately 85% of cases and may be distinguished only by this feature. The cellular variant of dermatofibroma may, however, shows focal CD34 positivity at the tumor periphery. Factor XIIIa can also aid in differentiation, as it is typically negative in dermatofibrosarcoma protuberans.

Dermatofibromas of large plaque-like sizes might be a kind of giant dermatofibromas. To be exact, very few cases of such instances have been reported, which are defined by the presence of large, tough plaques that can develop either on their own or as a result of some trauma like an insect bite[2,3,5]. Dermatofibroma (DF) is rarely found on the face, although the literature has documented a few such cases[9]. Just at the beginning of the millennium, Mentzel et al. in 2001 defined more than thirty thousand cases of dermatofibroma and among them, only 34 cases were referred to as being in the facial area composed of the forehead, ear, cheek, eyebrow, and nose. Out of these, the majority (17 cases) showed very aggressive behavior which led to the invasion of soft tissue and muscle. Histologically, only nine cases could be classified under the distinctly typical storiform pattern, while most were positive for actin spindle-shaped myofibroblasts and made up of cellular fascicles. In the same year, Estela et al. documented 22 instances of dermatofibroma occurring in the facial region over a span of twenty years, observing no unusual features. Involvement of deep tissue was seen in only three cases[10]. Atrophic dermatofibroma is an uncommon type of dermatofibroma that was first identified in 1987. It represents about 2% of all types of dermatofibromas[11]. This variant of dermatofibroma is most often seen in middle-aged women (40–50 years) and is localized to the upper trunk and upper arms. When diagnosing inwardly puckered, depressed lesions, this variant of dermatofibroma should be considered. Only a handful of cases of atrophic dermatofibroma have been reported in the literature so far[11].

The diagnosis is mainly clinical; however, in the case of atypical presentations like this, the histopathological examination together with IHC becomes the mainstay for accurate diagnosis and ruling out other diseases. Even though it looks otherwise, it always has a benign course and, in most cases, simple excision is sufficient for curing the patient. So far, there have been no reports of recurrences after excision.

Comparison with previously reported cases is informative. AlQusayer et al. described a facial dermatofibroma presenting as a small nodular lesion on the cheek of an adult patient[4]. Whereas the present case is distinguished by a much larger 8 × 4 cm atrophic plaque-like morphology rather than a discrete nodule. Iqbal and Mudaliar reported a 7 cm giant dermatofibroma on the leg of a 29-year-old male presenting as a non-ulcerated scaly plaque[7]. Our case is similar in its plaque-like configuration but differs in two important respects—facial location and the predominantly atrophic, depressed surface rather than a raised plaque. Cavallo et al. recently described a 20 cm plaque-like dermatofibroma on the antecubital region with peripheral satellite lesions, again on an extremity[8].

Atrophic dermatofibroma, first described in 1987, has classically been reported on the upper trunk and arms in middle-aged women; Alzaidieen et al. reported one such case on the leg of a 44-year-old woman in 2025[9]. To our knowledge, the combination of giant plaque-like size, atrophic morphology, and facial location has not previously been reported. Compared with the historical series of Mentzel et al., in which 17 of 34 facial dermatofibromas demonstrated aggressive behaviour with soft-tissue and muscle invasion[4], our case behaved as a typical benign fibrous histiocytoma on histopathology and immunohistochemistry, with no evidence of deep invasion. The CD68 and ASMA positivity with CD34 and S100 negativity in our case. is concordant with the immunoprofile reported across the cited literature and supports exclusion of dermatofibrosarcoma protuberans.

LIMITATIONS

This report describes a single case and therefore cannot establish the prevalence, natural history, or recurrence rate of this atypical facial variant of dermatofibroma. Dermoscopic evaluation, which has been increasingly used as an adjunct in the assessment of fibrohistiocytic tumours, was not performed on our patient. Additional immunohistochemical markers such as factor XIIIa, which can further help differentiate dermatofibroma

from dermatofibrosarcoma protuberans, were not available in our setting. Long-term follow-up beyond the immediate post-operative period is also limited. Larger case series and prospective registries, focused on atypical and facial dermatofibromas, are needed to better characterize the spectrum and behaviour of this entity.

CONCLUSION

This case report describes an unusual presentation of dermatofibroma as a giant plaque-like atrophic lesion on the face, an uncommon site for this tumor. Although the clinical morphology was atypical and closely simulated several inflammatory and connective-tissue dermatoses, histopathological examination and immunohistochemistry confirmed the features of the typical fibrous/fibrocellular variant of dermatofibroma. The diagnosis of such atypical lesions is challenging and mandates careful clinicopathological correlation, with histopathology and immunohistochemistry serving as the cornerstone for precise confirmation. Awareness of this variant among dermatologists, primary-care physicians, and pathologists is important to include dermatofibroma in the differential diagnosis of atypical facial plaques, to avoid misdiagnosis as morphea, lupus panniculitis, or dermatofibrosarcoma protuberans, and to guide timely surgical management with reassurance regarding the benign course and low recurrence risk.

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Authors' Contribution: FAK worked on methodology and writing the original draft. **SAA** conceptualized, supervised, and guided. **MS** validated, reviewed, and edited the draft. **NK** worked on data curation.

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