Keloidal Dermatofibroma of the Face: A Rare Series of Three Cases

SUDHA SRINIVASAN¹, FEME A MUNIRAJ, VIJAYASHREE RAGHAVAN¹

ABSTRACT
Dermatofibroma, also known as benign fibrous histiocytoma, is a cutaneous lesion that commonly affects young or middle-aged adults, with a slight preponderance in females. It usually presents as a single or multiple skin-coloured to reddish-brown nodule, primarily affecting the limbs. In the present case series, the authors reported three cases (two males and one female) of the keloidal variant of dermatofibroma, a benign fibrohistiocytic tumour. These cases involved patients aged between 32 and 55 years. All patients presented with nodular lesions on their faces, clinically diagnosed as dermatofibroma. Upon excision, the tumours exhibited circumscribed features with keloid-like areas, multinucleated giant cells, and inflammatory infiltrate. Keloidal dermatofibroma should be considered as a differential diagnosis when encountering tumours with keloid-like characteristics, especially in cases where incomplete excision may lead to recurrence. The present case series highlights the rarity of all three cases occurring on the face, as the prevalence of keloidal dermatofibroma is only 1% among other variants.

INTRODUCTION
Dermatofibroma is a benign soft tissue tumour that commonly affects middle-aged and young adults. It is usually easy to diagnose when it presents with classical features. While dermatofibroma typically occurs on the limbs, it rarely manifests on the face. Among the various variants of dermatofibroma, the keloidal type is particularly rare, with a prevalence of only 1% compared to other variants [1]. Additionally, its occurrence on the face is infrequent [2]. In the index case series report, the authors presented three cases of keloidal dermatofibroma on the face, highlighting their rarity.

Case 1
A 33-year-old female patient presented to the surgery clinic with a complaint of swelling near the left eyebrow that had been present for five years. On examination, a non tender swelling measuring 3 x 3 cm was observed near the left eyebrow, clinically diagnosed as an angular dermoid cyst. The differential diagnoses considered were dermatofibroma, sebaceous cyst, and cutaneous leiomyoma. There was no previous history of trauma or surgery. The lesion was completely excised under local anesthesia and sent for histopathological examination. The histopathological examination revealed a well-circumscribed tumour composed of spindle-shaped tumour cells arranged in a storiform pattern, along with extensive collagen, multinucleated giant cells, and haemosiderin-laden macrophages. Masson’s trichrome stain confirmed the presence of collagen [Table/Fig-1a-c]. Based on the histopathological examination, the case was diagnosed as keloidal dermatofibroma. No recurrence was noted during the three-year follow-up period.

Case 2
A 52-year-old male patient presented to the outpatient clinic with a complaint of a swelling in the cheek that had been present for three months. Clinically, it was diagnosed as keloid. Upon examination, a non tender, nodular, and hard swelling measuring 2×2 cm was observed in the cheek. The patient had a previous history of trauma. Based on the clinical examination, the differential diagnoses considered were dermatofibroma, nodular fasciitis, and basal cell carcinoma. The lesion was completely excised under local anesthesia and sent for histopathological examination. The histopathological examination revealed a well-circumscribed tumour composed of spindle-shaped tumour cells arranged in a storiform pattern, along with extensive collagen and an inflammatory infiltrate consisting of lymphocytes and plasma cells. Masson’s trichrome stain confirmed the presence of collagen [Table/Fig-2a-c]. Based on the histopathological examination, the case was diagnosed as keloidal dermatofibroma. No recurrence was noted during the two-year follow-up period.

Keywords: Dermal tumour, Recurrence, Soft tissue tumour, Variants

[Table/Fig-1]: a) Circumscribed nodular lesion in dermis, firm in consistency; b) Shows spindle shaped tumour cells in storiform pattern with interspersed collagen and multinucleated giant cells {Haematoxylin and Eosin (H&E) 40X}; c) Masson’s trichrome stain shows the presence of collagen {blue in colour (40X)}.

[Table/Fig-2]: a) Gross-dermal nodular lesion; b) Shows spindle shaped tumour cells with extensive hyalinised collagen, scattered inflammatory cells {Haematoxylin and Eosin (H&E) 40X}; c) Collagen stained blue in Masson’s trichrome stain (40X).
Case 3
A 35-year-old male patient presented to the outpatient clinic with a complaint of swelling on the forehead that had been present for seven years. It was clinically diagnosed as nodular fasciitis. Upon examination, a mobile swelling measuring 1.5×1.5 cm was observed with a firm consistency. There was no history of trauma or surgery. The differential diagnoses considered were dermatofibroma, calcified sebaceous cyst, and cutaneous leiomyoma. The lesion was completely excised under local anesthesia and sent for histopathological examination. The histopathological examination revealed a well-circumscribed tumour composed of spindle-shaped tumour cells with extensive collagen and a perivascular inflammatory infiltrate consisting of lymphocytes and plasma cells [Table/Fig-3a-c]. Based on the histopathological examination, the case was diagnosed as keloidal dermatofibroma. No recurrence was noted during the two-year follow-up period.

DISCUSSION
Dermatofibroma, also known as benign fibrous histiocytoma, is a cutaneous lesion that commonly affects young or middle-aged adults, with a slight preponderance for females [2]. It usually presents as a single or multiple skin-coloured to reddish-brown nodules, primarily affecting the limbs [3,4]. The size of the tumour can range from a few millimeters to 3 cm, and the colour may vary from light brown to red or black [2]. Diagnosis of dermatofibroma is straightforward when classical features are evident. Histologically, dermatofibromas are characterised by a well-circumscribed dermal tumour with epidermal hyperplasia and basal layer hyperpigmentation [5]. Proliferation of spindle cells, histiocytes, Touton giant cells, and the presence of lipidised siderophages are also typical features. Cytological atypia, pleomorphism, and mitotic activity may vary. Collagen trapping may be present.

Several variants of dermatofibroma have been described, including atrophic, keloidal, granular cell, myxoid, lichenoid, balloon cell, aneurysmal, haemodisectorial, cellular, epithelioid, atypical, lipiodised, clear cell, palisading, and signet-ring cell variants [6-12]. Among these variants, keloidal dermatofibroma accounts for only 1% of cases [1]. Accurately diagnosing these variants is crucial to prevent misdiagnosis as an aggressive neoplasm. Keloidal dermatofibroma is rare and seldom occurs on the face [2]. The occurrence of keloidal dermatofibroma on the face has been discussed in previous literature, and the present case series summarises the findings, as shown in [Table/Fig-4].

Dermatofibroma is often develops after trauma. Clinical differential diagnoses may include epidermal cysts, dermatofibrosarcoma protuberans, nodular fasciitis, cutaneous leiomyoma, basal cell carcinoma, and keloids. In keloids, fibroblasts are arranged without any organisation [13]. Epidermal cysts exhibit stratified squamous epithelium with keratin. Dermatofibrosarcoma protuberans is an infiltrative tumour that extends into the subcutis without keloid-like collagen. Nodular fasciitis is a benign neoplasm composed of spindle cells in a storiform pattern, with the stroma showing myxoid changes, extravasated red blood cells, and inflammatory cells. Cutaneous leiomyoma exhibits spindle cells in a fascicular pattern without inflammatory cells or multinucleated cells. Basal cell carcinoma is composed of basaloid cells arranged in lobules with peripheral palisading and retraction artifact, and numerous mitotic figures.

Local recurrence is uncommon in classical dermatofibromas occurring on the lower extremities, even if the excision is incomplete. In contrast, incomplete excision of dermatofibromas on the face is associated with local recurrence. The treatment of choice for dermatofibromas on the face is wide local excision with a skin graft. Therefore, prompt diagnosis of keloidal dermatofibroma is necessary to prevent recurrence and patient discomfort [2,5,13].

CONCLUSION(S)
The authors have presented a case series of keloidal-type dermatofibroma due to its rarity. Clinicians often struggle in achieving a correct diagnosis of keloidal dermatofibroma. However, with a careful morphological approach and the use of connective tissue stains, arriving at an accurate diagnosis becomes easier. It has been reported that keloidal dermatofibroma may recur if not completely excised. However, in the present case series, the authors did not observe any recurrence during the follow-up period after complete excision. Nevertheless, considering the possibility of recurrence with incomplete excision, the authors would like to emphasise the importance of awareness of this entity among pathologists and surgeons.

REFERENCES
PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Pathology, Chettinad Hospital and Research Institute, Chettinad Academy and Research Institute, Kelambakkam, Tamil Nadu, India.
2. Professor, Department of Pathology, Chettinad Hospital and Research Institute, Chettinad Academy and Research Institute, Kelambakkam, Tamil Nadu, India.
3. Professor, Department of Pathology, Chettinad Hospital and Research Institute, Chettinad Academy and Research Institute, Kelambakkam, Tamil Nadu, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Femela Muniraj,
Professor, Department of Pathology, Chettinad Hospital and Research Institute, Kelambakkam-603103, Tamil Nadu, India.
E-mail: fppathology@gmail.com

AUTHOR DECLARATION:
Financial or Other Competing Interests: None
Was informed consent obtained from the subjects involved in the study? Yes
For any images presented appropriate consent has been obtained from the subjects. NA

PLAGIARISM CHECKING METHODS: [Jain H et al.]
• Plagiarism X-checker: Apr 25, 2023
• Manual Googling: Jun 27, 2023
• iThenticate Software: Jun 30, 2023 (14%)

ETYMOLOGY: Author Origin

EMENDATIONS:
Date of Submission: Apr 18, 2023
Date of Peer Review: May 30, 2023
Date of Acceptance: Jul 03, 2023
Date of Publishing: Oct 01, 2023


