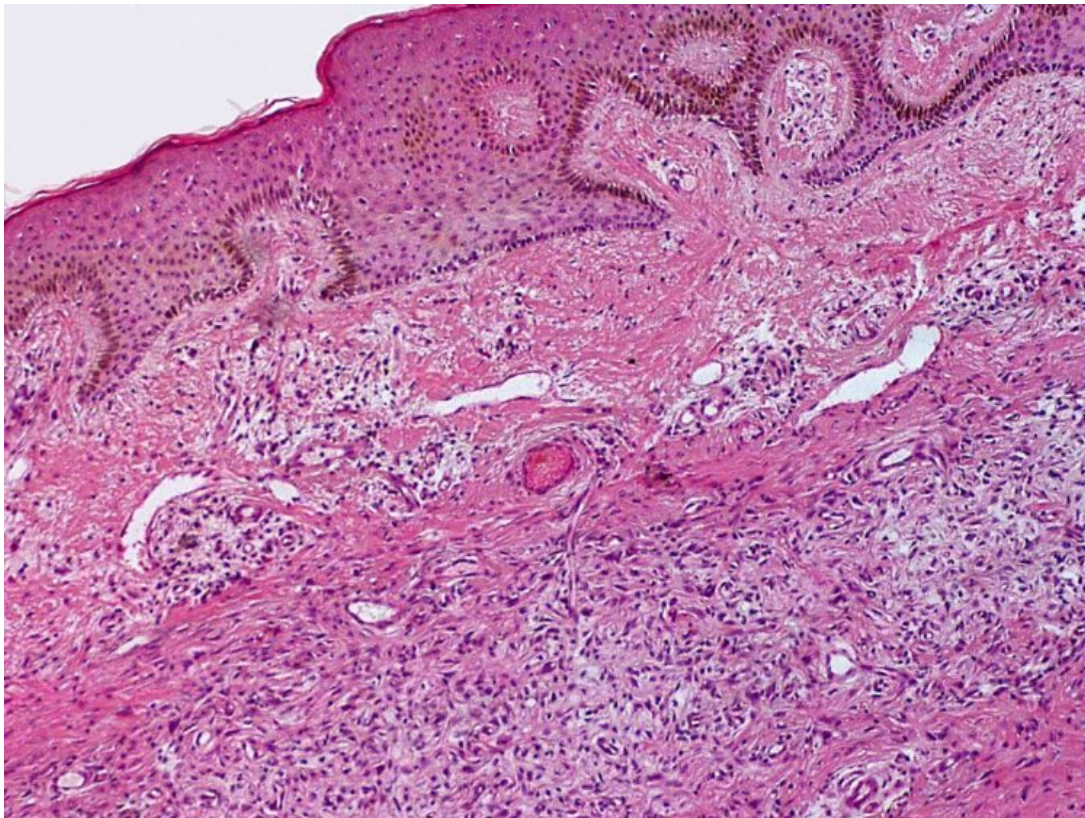


Dermatofibroma — Pathology Outlines and Treatment

[See online here](#)

Dermatofibroma, or benign fibrous histiocytoma, is a common cutaneous nodule that may be superficial or deep. It is the most common painful skin tumor and affects mostly women. Although the precise underlying etiology and mechanism for the development of this skin pathology are not well known, it is believed to be a neoplastic process owing to its clonal proliferation growth. While the patients may often be asymptomatic, the tumor growth may exhibit rapid growth or may remain static for several years. An intervention is not called for unless the patient is symptomatic.



Definition and Epidemiology of Dermatofibromas

Dermatofibroma is a **common mesenchymal growth of the skin** where skin fibroblasts are the major cell constituents. It is also known as **cutaneous benign fibrous histiocytoma** or **sclerosing hemangioma**.

A common mesenchymal tumor of the skin, dermatofibroma is known to have a **similar occurrence across all races**. There is a **predilection for women**, mostly occurring in

the lower legs, and in early adulthood, from **20** to **30** years of age. It can, however, occur at any age. The estimated female to male ratio of occurrence is 2.6 to 1. The average age at the time of presentation is 36.2 years.

Pathophysiology of Dermatofibromas

The exact mechanism for the development of a dermatofibroma is unknown, and clinicians are divided on whether it is a **reactive** process or a **neoplastic** one. Associations with **trauma** and **insect bites** have led some to believe that a reparative or reactive process may be the underlying pathology. However, recent demonstrations of **chromosomal abnormalities** and **clonal proliferation** have hinted more towards a neoplastic mechanism.

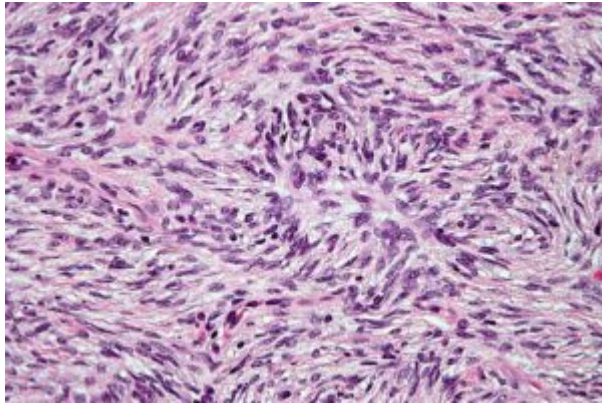
More likely, however, it is likely to arise from a distortion of **protein kinase C activity**. Immunohistochemical testing results have been found to be more consistent with the presence of **factor XIIIa** and **fascin** (dendritic cell markers) than with MAC - 387, a histiocyte marker.

Mast cells are found in abnormally high numbers in dermatofibroma. They release fibrogenic factors like transforming growth factor-beta (**TGF beta**) and trigger the fibrosis seen in dermatofibroma. In the **epithelioid and atypical dermatofibroma variants** specifically, **ALK gene rearrangement and overexpression** have been implicated.

Clinical Features of Dermatofibromas

- Measuring about **0.5-1 cm** in size, dermatofibromas are characteristically **firm, indurated, and mobile**.
- The lesions may be **solitary or multiple** (defined as at least 15 but most commonly benign), **generalized**, or **neoplastic**. Multiple lesions are usually associated with an immunosuppressive condition like **HIV, myasthenia gravis, systemic lupus erythematosus, diabetes**, etc.
- When neoplastic, they are usually confined to the **soles** and **palms** and may grow rapidly within weeks. However, some have been found to remain static for years.
- **Lateral compression** produces a dimple-like depression in the overlying skin which disappears with the release of pressure. This is called **button-holing** but is not specific to dermatofibroma. The positive buttonhole sign is also seen in type-1 neurofibromatosis (Von-Recklinghausen's disease) and anetoderma.
- Regression of these lesions may leave **hypopigmented spots**.
- Dermatofibromas arising on the **face** are quite uncommon but distinct from lesions seen at other sites. More commonly of the cellular type, they may extend into subcutaneous fat or muscle and recur frequently. This is why a wider initial excision is warranted in this case.
- **Local recurrence** is a feature of the atypical variant as well, along with **distant metastasis**.
- Up to 58% of the tumors are asymptomatic. 20.5% are tender to palpation and 11.5% are painful. The most common symptom, however, is pruritic sensation or itching.

Differential Diagnosis



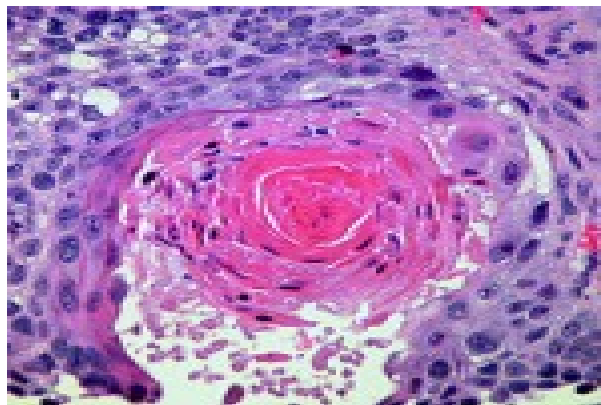
[Image](#): "Very high magnification micrograph of the storiform pattern in a case of dermatofibrosarcoma protuberans. H&E stain." By Nephron - Own work. License: [CC BY-SA 3.0](#)

The deep variant of dermatofibroma may be confused with an entity called **dermatofibrosarcoma protuberans** that is rather typically found on the trunk in younger adults. The size of this lesion is much larger, approximately 1–2cm.

In **ambiguous cases**, where the histological picture of the cellular variant closely resembles that of dermatofibrosarcoma protuberans, **immunolabeling** can be useful in distinguishing the factor XIIIa- and fascin-positive dermatofibroma from the latter (found to be negative for these markers).

However, the following skin pathologies are also to be considered in the differential:

- Atypical fibroxanthoma
- [Basal cell carcinoma](#)



[Image](#): "Squamous cell carcinoma." By Phil Schatz. License: [CC BY 4.0](#)

- Juvenile xanthogranuloma
- [Leiomyoma](#)
- [Melanoma](#)
- Nevus
- Pilomatrixoma
- Prurigo nodularis scar
- [Squamous cell carcinoma](#)
- Neurilemmoma

Investigations for Dermatofibroma

While the diagnosis of dermatofibroma is usually made **clinically**, confirmation can be undertaken by histological analysis on biopsy.

Biopsy

Histological pictures obtained on punch biopsy of the lesion reveal information on not just the type of variant but also help in making crucial decisions regarding management.

A typical picture that is most often encountered is:

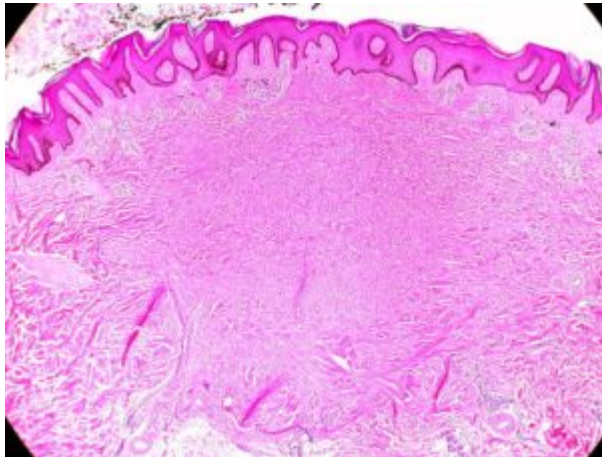


Image: "Histopathology of dermatofibroma." By Ed Uthman from Houston, TX, USA. Uploaded by CFCF. License: [CC BY 2.0](https://creativecommons.org/licenses/by/2.0/)

- **Epidermis:** usually hyperplastic and hyperpigmented, known as the dirty fingernail sign
- **Dermis:** haphazardly arranged fascicles of plump, spindle-shaped cells that typically lack atypia
- **Edges of lesion:** not well defined, with individual cells infiltrating between thickened and hyalinized collagen bundles (typical keloidal picture); also known as '**collagen trapping**'.
- **Mitotic figures** may be present, but atypia is characteristically not seen.

A **cellular** variant shows a **storiform** arrangement embedded in the deep dermis, with a much larger size and increased cellularity. It may often extend into the superficial subcutaneous tissue making it difficult to distinguish from an entity called dermatofibrosarcoma protuberans.

A rare form of **atypical** variant shows atypical features against a background of the classic picture of dermatofibroma. **Pleomorphic, spindle-shaped, plump, or polyhedral-shaped cells** are seen with **keloidal collagen** and **storiform** arrangement. The histopathological classification of dermatofibromas is given below:

- Fibrocollagenous (40.1%)
- Histiocytic (13.1%)
- Cellular (11.5%)
- Aneurysmal (7.4%)
- Angiomatous (6.5%)
- Sclerotic (6.5%)

- Other (14.9%)

Dermatoscopy

Visualization under a dermatoscope usually presents a **peripheral pigment network** with a **central white area**.

Serology

Indicated in cases where **immunosuppressive conditions** are suspected.

Management of Dermatofibroma

An intervention is usually not indicated in **asymptomatic** cases, which constitute the majority. **Watchful waiting** and **reassurance** suffice.

For **symptomatic cases**:

- **Medical management**: This is for patients who do not desire to undergo a surgical procedure. Topical or intralesional **corticosteroids** may help relieve the symptoms.
- **Cryosurgery** is another option to flatten out the dermatofibroma but is usually not curative.
- **Surgical management** includes cutaneous shaving, excisional surgery, and ablative laser surgery. This is usually indicated for cases where repeated trauma is suffered due to shaving on the lower legs. 20% of the cellular variants are known to recur after a simple biopsy and thus warrant a **fusiform excision** into the superficial subcutaneous tissue. For deeper variants, **excision to the deeper subcutaneous tissue** is done to decrease the risk for recurrence.

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