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Dermatofibrosarcoma protuberans

What is dermatofibrosarcoma protuberans (DFSP)?

DFSP is an uncommon skin tumour arising in the deeper layer of the skin (the dermis). It grows slowly but has a tendency to recur after excision. Luckily, it rarely spreads to other sites beyond the skin.

What causes DFSP?

The cause is unknown, but an injury to the affected skin may be a predisposing factor. Recent advances show abnormalities of chromosomes within the tumour cells, which may account for their unrestrained growth compared to the surrounding normal cells.

Who is at risk of DFSP?

It usually presents in early or middle adult life between 20 and 50 years of age, but all ages can be affected. The tumour is rare in children. Males are affected slightly more frequently than females. There does not appear to be any racial predilection.

What are the signs and symptoms of DFSP?

DFSP usually presents as a painless thickened area of skin (plaque) and/or nodule that feels rubbery or firm to touch and is fixed to the underlying skin. It may be red-brown or skin coloured. It usually grows very slowly over months to years.

Rarely it presents as a soft depressed area of skin making the diagnosis even more difficult. DFSP may range in size from 0.5 to 25 cm in diameter. Fifty to sixty percent of tumours arise on the trunk often in the shoulder and chest area. The remaining 35% of tumours are found on the limbs and 10 to 15% on the head and neck region.

DFSP is often diagnosed when it enters a more rapid growth phase giving rise to larger lesions. Neglected tumours may reach large proportions.

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How is DFSP diagnosed?

The absence of symptoms often leads to a delay in diagnosis. Redness and pain only occur in 15% of cases. It is often mistaken for other skin conditions particularly in the early stages. [Skin biopsy](#) is needed to confirm the diagnosis.

It has a characteristic appearance under the microscope with densely arranged spindle shaped cells. It may be difficult to assess complete removal.

What is the treatment for DFSP?

Treatment consists of wide excision of the lesion with several millimetres of normal skin around the margins.

Mohs micrographic surgery, which is a special surgical technique to control tumour margins, is sometimes used to check that all the abnormal cells have been excised.

[Radiotherapy](#) is sometimes used in addition to surgery. Chemotherapy is ineffective.

What is the prognosis?

The tumour only metastasises (spread beyond the skin) in 5% of cases. It spreads in 1% via lymphatic vessels to the regional lymph glands and in 4% via the blood stream, most commonly to the lung followed by the brain, bone and heart.

Local recurrences arise in 11–20% of cases, usually within 3 years of initial surgery, so follow-up is important.

Related information

On DermNet NZ:

- [Dermatofibroma](#)

Other websites:

- [Dermatofibrosarcoma Protuberans](#) - emedicine dermatology, the online textbook

Books about skin diseases:

See the [DermNet NZ bookstore](#)

Author: Dr Vania Sinovich MBChB FRACP, Dept of Dermatology Health Waikato

DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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