



DERMATOFIBROSARCOMA PROTUBERANS (DFSP)

What are the aims of this leaflet?

This leaflet has been written to help you understand more about dermatofibrosarcoma protuberans. It tells you about what it is, what causes it, what can be done about it and where you can find out more about it.

What is dermatofibrosarcoma protuberans?

Dermatofibrosarcoma protuberans is a very rare type of skin cancer. It usually occurs on the trunk of the body, for example the chest and shoulders, however it can also occur on the limbs and in some cases on the head and neck. It is thought to develop in the deep layer of the skin (the dermis) and can invade deeper tissue such as fat and muscle. Although it grows slowly, it can be quite extensive and can have a high recurrence rate even if removed by surgery. Dermatofibrosarcoma protuberans can, however, be cured if completely removed with a wide margin of normal tissue. Dermatofibrosarcoma protuberans only very rarely spreads to other parts of the body.

What causes dermatofibrosarcoma protuberans?

It is not clear what causes this type of skin cancer. It is thought that trauma or injury to the skin may be predisposing factors.

Is dermatofibrosarcoma protuberans hereditary?

There is no evidence to suggest that dermatofibrosarcoma protuberans is hereditary. It occurs in people of all races and ages, however evidence suggests that it is most likely to occur in people between the ages of 20 and 50.

What do dermatofibrosarcoma protuberans look like?

Dermatofibrosarcoma protuberans is usually slow growing, painless and appears as a thickened area of skin and/or lump which is attached to the underlying skin. It can range in colour from flesh coloured to pinkish/brown and occasionally can even be bluish. It can also present as a soft indented area on the skin which can make diagnosis difficult. If left for several years the dermatofibrosarcoma protuberans can ulcerate.

What are the symptoms of dermatofibrosarcoma protuberans?

The symptoms of a dermatofibrosarcoma protuberans are usually very subtle, if any at all. The patient may notice a thickened/discoloured patch of skin, an indented area or a lump increasing in size. A dermatofibrosarcoma protuberans is generally painless, so it is usually a change in size or colour that alerts the person to their presence.

How is dermatofibrosarcoma protuberans diagnosed?

Due to its rarity, the diagnosis is often delayed. It may be mistaken for a number of other skin conditions. If dermatofibrosarcoma protuberans is suspected, a piece of abnormal skin will be removed and tested (biopsied) to confirm the diagnosis.

Can dermatofibrosarcoma protuberans be cured?

Dermatofibrosarcoma protuberans can be cured if completely removed. However, it can recur even after surgical intervention. Regular follow-ups post-treatment are therefore required to monitor the skin.

How can dermatofibrosarcoma protuberans be treated?

Surgical excision (removal) is the most common and effective treatment for dermatofibrosarcoma protuberans. There are two main types of surgical procedure used:

A wide excision . this involves removal of the lesion with an extra margin of normal skin around the edges to ensure no abnormal cells are left behind. The wound will then be reconstructed (closed) using the most suitable method. This may happen under local or general anaesthetic depending on the size and location of the dermatofibrosarcoma protuberans.

Mohs' micrographic surgery . this is a specialised surgical technique (usually performed by a dermatologist) whereby the dermatofibrosarcoma protuberans is surgically removed in stages under local anaesthetic. The abnormal tissue will be removed and examined under the microscope overnight. A dressing is placed over the wound until the results are ready, and if there are any remaining abnormal cells this process is repeated until the dermatofibrosarcoma protuberans has been completely removed. The wound will then either be repaired by the dermatologist or a plastic surgeon, depending on the size and location of the wound.

Extremely rarely, dermatofibrosarcoma protuberans may spread to other parts of the body. If this has occurred, further treatment may be required such as radiotherapy or chemotherapy.

Self care (what can I do?)

Due to the subtle clinical appearance of dermatofibrosarcoma protuberans, it can be difficult for even health professionals to detect. It is advisable that you do regular checks of your scar at home and if you have any suspicious areas or changes in the scar you should contact your GP or dermatologist.

Where can I get more information about dermatofibrosarcoma protuberans?

Links to patient support groups:

Macmillan Cancer Support

89 Albert Embankment

London

SE1 7UQ

Helpline (for emotional support): 0808 808 2020

Helpline (for information): 0808 800 1234

Website: www.macmillan.org.uk

The Rarer Cancers Foundation

The Great Barn

Godmersham Park

Canterbury

Kent, CT4 7DT

Tel: 0122 773 8279

Helpline: 0800 334 5551

Email: helpline@rarercancers.org.uk

Website: <http://www.rarercancers.org.uk>

Web links to detailed leaflets:

<http://emedicine.medscape.com/article/1100203-overview>

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: its contents, however, may occasionally differ from the advice given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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