PATIENT INFORMATION LEAFLET

DERMATOFIBROSARCOMA PROTUBERANS

WHAT ARE THE AIMS OF THIS LEAFLET?

This leaflet has been written to help you understand more about dermatofibrosarcoma protuberans (DFSP). 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?

DFSP is a very rare type of soft tissue cancer. It usually occurs on the trunk, often the chest and shoulders; however, it can also affect the limbs, head and neck and rarely the genitalia. It starts in the deep layer of the skin (the dermis) and can spread locally to surrounding structures such as fat and muscle. Although it grows very slowly, it can become quite large. It can often be completely removed surgically by including a margin of normal tissue, or with a specialized form of surgery called Mohs surgery. DFSP may come back in the same place even if completely removed by surgery (in 1-3%, or 1 to 3 out of every 100 people). It is very rare for DFSP to spread to other parts of the body (occurring in approximately 1-7% of cases, or 1 to 7 out of every 100 people).

WHAT CAUSES DERMATOFIBROSARCOMA PROTUBERANS?

Trauma or injury to the skin may be partly responsible in some cases. It is not known why or how the tumour occurs.

IS DERMATOFIBROSARCOMA PROTUBERANS HEREDITARY?

No. It occurs in people of all races and ages, usually in early adult life. There have been some genetic changes identified in the cells of DFSP tumours, but these are not inherited and cannot be passed on.

WHAT DOES DERMATOFIBROSARCOMA PROTUBERANS LOOK LIKE?

DFSP is usually a slow growing, painless, thickened lumpy area of skin. It can be skin coloured, pink/red, brown/black or occasionally blue/purple. It can also feel like a soft or hardened sunken area on the skin. If left for several years DFSP can grow through the top layer of the skin, producing an ulcer.

WHAT ARE THE SYMPTOMS OF DERMATOFIBROSARCOMA PROTUBERANS?

Most people with DFSP do not have any symptoms. They may notice thick skin, skin changing colour, a sunken area or a lump increasing in size.

HOW IS DERMATOFIBROSARCOMA PROTUBERANS DIAGNOSED?

As DFSP is rare it may take time for it to be diagnosed. It may be mistaken for common harmless skin conditions such as cysts, dermatofibromas or keloids. If DFSP is suspected, a piece of the abnormal skin will be removed under local anaesthetic (a biopsy) and examined under the microscope for specific features of the disease.

CAN DERMATOFIBROSARCOMA PROTUBERANS BE CURED?

Yes, DFSP can be completely treated in most people. In about 97-99% (97 to 99 out of 100 people) surgically removing the abnormal skin will permanently treat the DFSP. However, it can come back (in 1-3%, or 1 to 3 out of every 100 people).

Follow-up visits to the dermatology clinic are usually for the first 5 years, longer for some. If DFSP has spread to other parts of the body removal may not be possible.

HOW CAN DERMATOFIBROSARCOMA PROTUBERANS BE TREATED?

DFSP is treated by surgical excision (removal). There are two main types of surgery:

Wide excision – this involves removal of the DFSP with a margin of normal skin around the edges. The wound will then be closed using stitches typically, sometimes by a skin flap (moving local skin) or graft (moving skin from further away). This may happen under local or general anaesthetic depending on the size and location of the DFSP, and the choice of the patient and surgeon.

Mohs micrographic surgery – this is a type of surgery in which the surgeon tries to spare skin, done under local anaesthetic. The abnormal tissue is removed and examined under the microscope immediately. A dressing is placed over the wound until the results are ready, and this process is repeated until the DFSP has been completely removed (for example, the edges of the tissue taken away are clear of any disease). The wound will then be repaired, sometimes by a different surgeon depending on its size and location. This type of surgery offers a higher cure rate than wide excision as full removal is confirmed before the wound is closed.

Very rarely, DFSP may spread to other parts of the body. Further treatment may then be required, such as radiotherapy or drugs. Radiotherapy or drugs (see below) can also be used if the DFSP cannot be removed completely by surgery.

DRUG TREATMENT FOR DERMATOFIBROSARCOMA PROTUBERANS

Imatinib is a chemotherapy medicine used for DFSP that cannot be removed with an operation or has spread within the body (a metastatic DFSP). It is taken as a tablet. Some patients may then be able to proceed to surgery if their DFSP shrinks.

WHAT TESTS DO I NEED?

Most people only need surgery for DFSP. If there are concerns that it may have spread, you may need X-rays, ultrasounds, MRI or CT scans. If any lumps develop in the scar, the lump will need to be biopsied (tissue samples taken) for examination under the microscope.

SELF-CARE (WHAT CAN I DO?)

DFSP can be difficult even for doctors to detect. After having had a DFSP, you should do regular checks of your scar at home; if you notice any changes in the scar, such as new lumps which do not settle within a week or two, you should contact your primary care physician, dermatologist or department leading your care.

WHERE CAN I GET MORE INFORMATION ABOUT DERMATOFIBROSARCOMA PROTUBERANS?

Links to patient support groups:

Macmillan Cancer Support

Helpline (for information): 0808 808 00 00

Website: https://www.macmillan.org.uk/

Web links to detailed information:

http://www.pcds.org.uk/clinical-guidance/dermatofibrosarcoma-protuberans

http://www.dermnetnz.org/lesions/dfsp.html

https://www.aad.org/public/diseases/skin-cancer/types/common/dfsp

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: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

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

BRITISH ASSOCIATION OF DERMATOLOGISTS

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