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 what it is, what causes it, what can be done about it and where you can find more information.

WHAT IS DERMATOFIBROSARCOMA PROTUBERANS?

DFSP is a very rare type of soft tissue cancer. Each year out of every million people only about 3 to 4 will get this condition. Cancer happens when cells in the body grow out of control.

DFSP affects soft tissue (flesh). This means areas of the body surrounding the organs and bones, including muscles, skin, and fat. It usually appears on the upper body (trunk), often the chest and shoulders. However, it can also affect the arms and legs, head and neck, and rarely the private parts (genitalia).

DFSP starts in the middle layer of the skin (called the dermis) and can spread to other parts of the area around it, such as fat and muscle. Although it grows very slowly, it can become quite large.

Usually, it can be removed completely during surgery (an operation). Some of the area around it may also need to be removed to lower the risk of it coming back. Sometimes a special type of surgery is used called Mohs-micrographic-surgery.

It is possible that DFSP comes back in the same place, even if it was completely removed. In most people, DFSP does not spread to other parts of the body, such as lungs and bones. However, this does happen in about 1 of every 100 people who get this condition.

WHAT CAUSES DERMATOFIBROSARCOMA PROTUBERANS?

The cause of why and how DFSP tumours occur is unknown.

Injury to the skin, such as blunt trauma, injection sites and burns, may be partly responsible in some cases.

Specific genetic changes within the skin cells have also been found.

IS DERMATOFIBROSARCOMA PROTUBERANS HEREDITARY?

No.

WHAT DOES DERMATOFIBROSARCOMA PROTUBERANS FEEL AND LOOK LIKE?

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

DFSP is usually a slow growing, painless, thickened lumpy area of skin. Because of this, it can be mistaken for a thickened or keloid scar, a cyst or a dermatofibroma (a harmless growth following local trauma, such as an insect bite or a puncture wound from rose thorn).

The skin can be normal in colour, pink/red, brown/black or sometimes blue/purple. The area of skin can also feel softer, or harder and sunken (lowered). If nothing is done about it for a long time (several years), DFSP can grow through the top layer of the skin and make an open wound (ulcer).

HOW IS DERMATOFIBROSARCOMA PROTUBERANS DIAGNOSED?

As DFSP is rare it may take time for it to be diagnosed. If your healthcare professional suspects it may be DFSP, a biopsy may be taken. The skin will be numbed with anaesthetic, and a small piece of skin will be taken to be analysed in the laboratory.

CAN DERMATOFIBROSARCOMA PROTUBERANS BE CURED?

Yes, DFSP can be completely treated in most people. In about 99% of cases (99 out of 100 people), DFSP can be completely removed with surgery. However, it is possible for it to come back. If this happens, more treatment, such as radiotherapy or other types of targeted oral treatment, may be needed.

Usually, you will be asked to come back to the dermatology clinic for at least 5 years. This is so the doctors can check it does not come back.

If DFSP spreads to other parts of the body, such as lungs and bones, it may not be possible to remove it .

HOW CAN DERMATOFIBROSARCOMA PROTUBERANS BE TREATED?

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

Wide excision – this type of surgery removes the DFSP tumour, plus an extra bit of tissue around the edge of it. The wound will then be closed using stitches, or sometimes by a skin flap (moving skin from beside the wound) or graft (moving skin from further away) to cover the area where skin has been surgically removed. This may happen under anaesthetic which can be:

- local (numbing the specific skin area only) or
- general anaesthetic (putting the patient in a controlled unconscious state).

The type of anaesthetic used depends 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 (operation) in which the surgeon tries to remove as little skin as possible. The local area of skin is numbed with anaesthetic. The abnormal tissue is then 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, there is no more disease on the edges of the tissue taken away). The wound will then be repaired, sometimes by a different surgeon depending on its size and location. The chances of fully removing the tumour are higher with this type of surgery, because the surgeon can check that it is fully removed before the wound is closed. However, this type of operation may not be available in all UK regions, because it requires a specialist.

Very rarely, DFSP may spread to other parts of the body. Further treatment may then be required, such as radiotherapy or other targeted treatments. These can also be used

if the DFSP cannot be removed completely by surgery.

WHAT TESTS DO I NEED?

Most people only need surgery for DFSP. If there are concerns that it may have spread to other parts of the body, your specialist might ask you to get one or more scans. These could be X-rays, ultrasounds, MRI or CT scans. If any more lumps develop in the scar, they will need to be looked at again by your doctor. It is possible that they may need to take another small piece of it (biopsy).

SELF-CARE (WHAT CAN I DO?)

DFSP can be difficult to detect, even for healthcare professionals. After having had a DFSP, you should regularly check your scar at home. If you see or feel any changes in the scar, such as new lumps which do not go back to normal within a week or two, you should contact your healthcare professional leading your care.

WHERE CAN I GET MORE INFORMATION ABOUT DERMATOFIBROSARCOMA PROTUBERANS?

Patient support groups providing information:

Macmillan Cancer Support
Web: https://www.macmillan.org.uk/
Web links to other relevant sources:
Sarcoma UK: https://sarcoma.org.uk/about-sarcoma/what-is-sarcoma/types-of-sarcoma/dermatofibrosarcoma-protuberans-dfsp/
DermNetNZ:
www.dermnetnz.org/lesions/dfsp.html
AAD: www.aad.org/public/diseases/skin-cancer/types/common/dfsp

Please note that the BAD provides web links to additional resources to help people access a range of information about their treatment or skin condition. The views expressed in these external resources may not be shared by the BAD or its members. The BAD has no control of and does not endorse the content of external links.

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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