Understanding Dermatofibrosarcoma Protuberans: A Guide for Patients

Megan Hoang

Dermatofibrosarcoma protuberans (DFSP) is a rare skin cancer that affects about 4 out of 1 million people worldwide each year. DFSP is a sarcoma (a soft tissue tumor) that can involve the skin, fat, and rarely, muscle and bone. It accounts for 1-6% of all soft tissue sarcomas and 18% of all soft tissue sarcomas of the skin. DFSP begins in the middle layer of your skin, which is called the dermis.

At first, DFSP often looks like a rough, discolored patch of skin, a bruise, a small bump on the skin, or a pimple. It can also look like a birthmark in children or infants. As DFSP grows, multiple firm lumps of tissue can develop on your skin. These lumps may be violet to red-brown, become painful or tender, bleed, or break open easily. DFSP tumors often appear on your chest, abdomen, back, shoulders, pelvis, arms, legs, head, or neck.

DFSP usually affects adults aged 30-50 years old, though it can also affect children, including some infants at birth. DFSP is also most common in black patients. During pregnancy, DFSP can also grow faster. A gene change that occurs after birth has been suggested as key to the development of DFSP in about 90% of patients. Other potential risk factors for DFSP include a skin injury or scars from causes like burns, radiation, or tattoos.

DFSP is non-aggressive, slow-growing, and rarely spreads, though there is a high chance of the cancer coming back. DFSP has a high survival rate with appropriate treatment to remove the tumor and lower the chances of the cancer coming back. DFSP is more likely to spread if you do not get treatment.

Your doctor will closely examine your skin and perform a skin biopsy by removing some of the tumor for testing to confirm a diagnosis of DFSP. Imaging tests such as an MRI may also be performed to determine the extent of the cancer before surgery. These tumors are usually treated through surgical removal, preferably with Mohs surgery. Mohs surgery involves repeatedly cutting out layers of cancerous skin and some of the healthy tissue around it until no more cancer cells are found. Another treatment for DFSP is wide local excision. Other treatments could include imatinib (a chemotherapy medication) or radiation therapy.

Following diagnosis and treatment for DFSP, you should check your skin regularly to look for any skin changes. If you see any new, unusual, changing, or bleeding growths or bumps on your skin, make an appointment with your doctor. Make regular visits to your doctor as well to get skin exams every 4 months for the first 3 years after treatment, and then annually if the DFSP does not return.

Understanding Dermatofibrosarcoma Protuberans: A Guide for Caregivers

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At first, DFSP often looks like a rough, discolored patch of skin, a bruise, a small bump on the skin, or a pimple. It can also look like a birthmark in children or infants. As DFSP grows, multiple firm lumps of tissue can develop on your loved one's skin. These lumps may be violet to red-brown, become painful or tender, bleed, or break open easily. DFSP tumors often appear on your loved one's chest, abdomen, back, shoulders, pelvis, arms, legs, head, or neck.

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Following diagnosis and treatment for DFSP, you should help check your loved one's skin regularly to look for any skin changes. If there are any new, unusual, changing, or bleeding growths or bumps on your loved one's skin, make an appointment with their doctor. Make regular visits to their doctor as well to get skin exams every 4 months for the first 3 years after treatment, and then annually if the DFSP does not return.

References

1. Brooks J, Ramsey ML. Dermatofibrosarcoma Protuberans. 2023 Apr 14. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan–.
2. Gloster HM Jr. Dermatofibrosarcoma protuberans. J Am Acad Dermatol. 1996 Sep;35(3 Pt 1):355-74; quiz 375-6. Erratum in: J Am Acad Dermatol 1997 Apr;36(4):526.
3. Llombart B, Serra C, Requena C, Alsina M, Morgado-Carrasco D, Través V, Sanmartín O. Guidelines for Diagnosis and Treatment of Cutaneous Sarcomas: Dermatofibrosarcoma Protuberans. Actas Dermosifiliogr (Engl Ed). 2018 Dec;109(10):868-877. English, Spanish. Epub 2018 Jul 4.
4. National Comprehensive Cancer Network Dermatofibrosarcoma Protuberans and Merkel Cell Carcinoma. Dermatofibrosarcoma protuberans. Clinical practice guidelines in oncology. J Natl Compr Canc Netw. 2004 Jan;2(1):74-8.