DERMATOFIBROSARCOMA PROTUBERANS

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Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive soft tissue tumor which typically presents on the proximal extremities and trunk. It is the most common type of skin sarcoma. The cause of DFSP is not clearly understood but sometimes a history of trauma may be present. The lesion typically occurs in middle aged individuals. Males are slightly affected more commonly. The lesion has a low chance of metastasizing to other parts of the body although there have been reports.

The lesion appears as a smooth, firm, mobile, nodular raised area of skin that can measure up to a few centimeters. It can be flesh or red, purple to brown color. The borders are asymmetrical and the surface is very irregular and may begin indurated in the skin. To diagnose DFSP, a biopsy should be conducted. The tumor grows very slowly causing for a delayed diagnosis, of sometimes months to years.

Although the newest literature states that the treatment of choice for DFSP is Mohs surgery, it is not available in all places and not all Mohs surgeons feel comfortable operating on this type of tumor. Mohs surgery allows less tissue removal and more adequate margin assessment. There are several other options for treating DFSP. Previously, surgical excision with generous margins (2-3cm) was the treatment of choice. Moreover, radiation is sometimes given in addition to surgery to prevent recurrence. Radiation can also be used if surgical excision significantly affects patients cosmetically. The drug Imatinib is another option for non-surgical candidates or when DFSP is unresectable, recurrent or metastatic. After treatment, patients must be seen every six months for three years due to the high recurrence rate. In cases of metastases, a multidisciplinary approach should be undertaken including a dermatologist, surgical and medical oncologist.