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Dermatofibrosarcoma Protuberans

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive soft tissue sarcoma that arises in the dermis and subcutaneous tissues. It is the most common form of cutaneous soft tissue sarcoma and is characterized by a slow-growing, indolent tumor that typically manifests in the proximal extremities and trunk. Although DFSP is generally considered a low-grade malignancy with a low risk of metastasis, it has a tendency to recur locally, making appropriate management critical. This neoplasm most often affects middle-aged individuals, with a slight male predominance. The exact etiology of DFSP remains poorly understood, though a history of trauma has been reported in some cases.

Clinical Presentation

DFSP typically presents as a smooth, firm, mobile, nodular lesion that is raised above the surrounding skin. The lesion can range in size from a few centimeters to several inches and can vary in color from flesh-toned to red, purple, or brown. The borders of the tumor are often asymmetrical, with an irregular surface that may become indurated over time. Because of its slow growth, DFSP is often diagnosed months to years after the initial onset of symptoms, and patients may not notice the tumor until it has reached a significant size. Although rare, reports of metastasis to distant organs have been documented, particularly in cases of recurrent or aggressive DFSP.

Diagnosis

Diagnosis of DFSP is confirmed through a skin biopsy, which typically reveals a spindle cell tumor with storiform patterns of growth. Immunohistochemistry may be used to identify the tumor's characteristic markers, such as CD34, which is expressed in the majority of DFSP lesions. Imaging studies, such as MRI, can help assess the extent of local involvement, which is critical for surgical planning. DFSP is known for its locally invasive growth pattern, often infiltrating surrounding tissues in a manner that is difficult to appreciate clinically.

Pathophysiology

DFSP arises from the dermis or subcutaneous tissue and is characterized by fibroblastic or myofibroblastic cell proliferation. The tumor is often associated with a chromosomal translocation, most commonly the t(17;22) translocation, which leads to the COL1A1-PDGFB fusion gene. This gene fusion results in the overproduction of platelet-derived growth factor B (PDGF-B), which stimulates the proliferation of fibroblasts and other mesenchymal cells, contributing to the tumor's aggressive behavior. The tumor typically exhibits slow growth but can lead to extensive local invasion if left untreated. Although metastasis is rare, it can occur in advanced stages, particularly when the tumor is left untreated or recurrent.

Treatment

The treatment of DFSP is primarily surgical. The goal of surgery is to achieve complete excision with adequate margins to minimize the risk of local recurrence. In the past, the standard surgical approach involved excision with generous 2–3 cm margins, but recent studies have emphasized the use of Mohs micrographic surgery (MMS) as the preferred treatment modality. Mohs surgery offers the advantage of real-time tissue margin assessment, which allows for maximal tumor removal while sparing healthy tissue, leading to better cosmetic outcomes and a reduced risk of recurrence. However, Mohs surgery is not available in all settings, and not all Mohs surgeons may be comfortable performing this procedure on DFSP tumors.

For large or recurrent DFSP tumors, radiation therapy may be used as an adjunct to surgery. Radiation can help reduce the risk of recurrence, especially in cases where the tumor is located in difficult-to-reach areas or when surgery might lead to significant cosmetic defects. In cases where surgical excision is not possible or when DFSP is metastatic or recurrent, systemic treatment options may be considered. One such option is Imatinib, a tyrosine kinase inhibitor that targets the PDGF receptor and has shown efficacy in treating DFSP, particularly in cases where surgical resection is not feasible. Imatinib is particularly useful in managing unresectable or recurrent DFSP and can serve as a viable alternative for patients who are not candidates for surgery.

Follow-Up and Prognosis

Due to the tendency of DFSP to recur locally, patients should be monitored closely after treatment. Follow-up visits are typically scheduled every six months for the first three years, with annual exams thereafter. This frequent follow-up is essential to detect any signs of recurrence at an early stage, as recurrent DFSP can often be treated successfully if caught early. The prognosis for patients with DFSP is generally favorable, particularly in cases where the tumor is detected early and completely excised. However, distant metastasis can occur in advanced stages, necessitating a multidisciplinary approach that includes dermatologists, surgical oncologists, and medical oncologists.

Conclusion

Dermatofibrosarcoma protuberans is a locally aggressive soft tissue tumor that requires early recognition and appropriate management. The mainstay of treatment is surgical excision, with Mohs micrographic surgery emerging as the preferred method due to its precision and ability to minimize tissue loss. Adjunctive therapies such as radiation therapy and Imatinib offer additional options for cases where surgery is not feasible or when recurrence occurs. Given the potential for recurrence, patients with DFSP must undergo regular follow-up to ensure the early detection of any recurrence or metastasis. With appropriate treatment, the prognosis for patients with DFSP is generally positive, though close monitoring is essential for long-term success.

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