Dermatofibrosarcoma protuberans

Dermatofibrosarcoma protuberans (DFSP) is a rare low-grade sarcoma of the fibroblast originating from the dermal layer of the skin, characterized by a locally aggressive growth and high rate of local recurrence.

Mutations in PDGFB are associated with meningioma. Reciprocal translocations between chromosome 22 and chromosome 17, at sites where this gene and that for COL1A1 are located, are associated with a particular type of skin tumor called dermatofibrosarcoma protuberans resulting from unregulated expression of growth factor. Two splice variants have been identified for this gene.

Outcome

A successful treatment and management depends on achieving local control and preventing cosmetic and functional deficit; all efforts should be made for complete excision. Postoperative follow-up recommended for highly suspicious cases and annual checkups should be performed up to 5 years after definitive therapy [1)]

Case reports

Two patients underwent a wide radical excision of recurrent scalp DFSP which was reconstructed with translational skin flap and split-thickness skin graft. We described above cases several years ago with a local excision of the tumor; recently, they developed local recurrence of DFSP with calvarial involvement. We then performed a wide radical excision, with craniectomy of the cranial defect followed by cranioplasty using titanium mesh, continuing with reconstruction.

A successful treatment and management depends on achieving local control and preventing cosmetic and functional deficit; all efforts should be made for complete excision. Postoperative follow-up recommended for highly suspicious cases and annual checkups should be performed up to 5 years after definitive therapy [2)]

Unclassified

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[1)] , [2)]
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