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Original Research Paper

General Surgery

A RARE CASE OF DERMATOFIBROSARCOMA PROTUBERANS OF UPPER LIMB

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ABSTRACT Dermatofibrosarcoma Protuberans Is A Rare Soft Tissue Sarcoma That Accounts For 0.1 % Of Cancers.most Commonly Occurs In Trunk And Extremities.less Than 1% Occur In Perineal Region.(1)it Is A Rare Soft Tissue Tumour That Involves Dermis Subcutaneous Fat And In Rare Cases Involve Muscles And Fascia.it Is A Intermediate Grade Malignancy With Low Likelihood To Metastatize And Has High Recurrence Rate Occuring In Middle Aged.

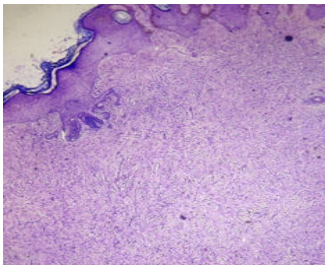
KEYWORDS : dermatofibrosarcoma, sarcoma,malignancy

Case Study:

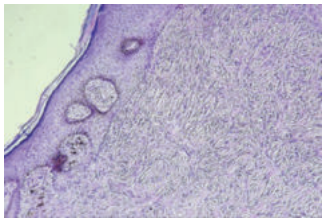
A Previously Healthy 39 Year Old Female Presented With Complaints Of Painless Exophytic Nodule Over Left Forearm With One Year Duration. On Examination A Nontender Purplish Nodule Of Size 1.5x1.5 Cm Noted In Left Forearm Which Is Irregular And Has Rough Surface. Fnac Was Inconclusive With Clinical Suspicion Of Neoplasm We Proceeded With Wide Local Excision With 2cm Margin Clearance. Postoperative Period Was Uneventful. Histopathology Report Showed A Hyperplastic Epithelium With With Underlying Poorly Circumscribed Neoplasm In Dermis Arranged As Nodule Composed Of Spindle Cells In Storiform Pattern Extending To Subcuticular Layer At Few Places. Cd34 Showed Positivity In 80% Of Tumour Cells. patient Is Currently Under Followup For 6 Months

DISCUSSION:

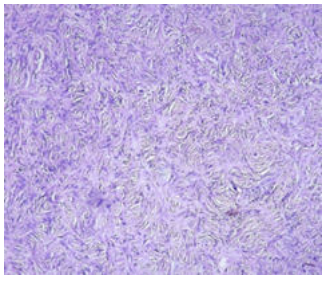
Dermatofibrosarcoma Protuberans(dfsp) Is A Malignant Soft Tissue Tumour . Its Indolent Growth Pattern Can Be Associated With Initial Miisdiagnosis Or Delayed Msdiagnosis And Treatment . Dfsp Is Associated With Translocation Of Genetic Material Between Chromosome 17 And 22 T(17;22) Fuses Part Of Col1a1 Gene Of 17 With Pdgfb Of Chromosome 22 .skin Biopsy Confirms Diiagnosis . Treatment Is Wide Local Excision With Meticulous Histologic Examination Of Specimen To Ensure Adequate Margins. reconstruction Or Tissue Underming Should Be Avoided Until Negative Histologic Margin Are Confirmed Imatinib A Protein Tyrosine Kinase Inhibitor Can Be Used For Recurrent Or Unresectable Dfsp With T(17;22)(2) Translocation. Radiation Theraphy Has Been Utilized For 10 Year Disease Free Survival And Could Be Employed If Disease Resection Is Not Faesible



Storiform Pattern



Fascicles Of Tumour Cells



Spindle Cell Morphology

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