Abstract:
Introduction - Plexiform neurofibroma or Pachydermatocoele is an uncommon variety of generalized neurofibromatosis, usually associated with Trigeminal nerve. We report two cases of plexiform neurofibroma. Case 1) 18-year-old girl with left forehead neurofibroma, Case 2) 14-year-old girl with right leg neurofibroma. Clinical Presentation - The neurofibroma started in childhood. Family history of neurofibroma was positive in both cases. Cafe' au lait spots, a useful diagnostic sign, seen in both patients. Intervention - Subtotal excision was done to correct cosmetic deformity. Conclusion - Surgical excision is the widely accepted form of treatment. Malignant changes to be ruled out by careful histopathological evaluation. Prognosis is usually good with early surgical treatment.

Keyword: Pachydermatocoele, Von Recklinghausen's disease, Watson's disease, Gadolinium Enhanced MRI

INTRODUCTION:
Plexiform neurofibroma or pachydermatocoele is a type of neurofibromatosis caused by excessive growth of the neural tissue in the subcutaneous fat. It is commonly seen in connection with the branches of trigeminal nerve. It has also been reported in retroperitoneal region, paraspinal and mediastinal area, anterior abdominal wall. We report two cases of plexiform neurofibromatosis.

CASE REPORT
CASE-1:
A 18-year-old girl with the history of swelling in her left forehead since childhood, which gradually increased in size in the past two years. It was painless and not obstructing the vision. Patient on examination had a multilobular swelling measuring 10cmx8cm on the left side of her forehead. It was extending into the medialcanthus region. Apart from routine investigations Gadolinium enhanced MRI was done (fig.1).
Patient was treated by subtotal excision through bicoronal incision. Post op: Patient had transient ptosis, which improved over time. Ptosis, probably due to post operative edema, redundant skin following tumor excision or Neuro praxia. Patient treated with steroids post operatively. Within a week patient recovered gradually.

**CASE: 2**

A 14 year old female with the complaints of swelling in the Right leg, posterior to ankle joint, on either side of Tendo Achilles(fig.5). Swelling started from age of 6. Present with size of 6x11 cm. Well defined nodular soft swelling. Cafe’- au- lait spots ,5-6 nodules, over chest & back present. Her father had multiple neuro fibromatous swellings over face, both upperlimbs & trunk.
DISCUSSION:
Plexiform neurofibromas are frequently associated with hypertrophy of the soft tissue and hyperpigmentation. Their growth can cause destruction or compression of local tissue, causing significant morbidity. Other names are Von Recklinghausen’s disease, Watson’s disease. It is an autosomal dominant genetic disorder of NF1 type. NF1 is caused by mutation in the neurofibromin(6)gene localised to band 11.2 of the long arm of chromosome 17.Chances of occurrence in affected family
is about 50% (6). **Common sites of origin:** The trunk - 43% to 44%. The extremities - 15% to 38%. The head and neck - 18% to 42% of patients (6). The incidence is 1 in 3000 live births (4). Craniofacial neurofibromas cause significant facial disfigurement. Plexiform neurofibromas have a high propensity for malignant degeneration (2) of 6 - 13%. Pain is the most reliable indicator (6) of malignant degeneration.

**1987 National Institute Of Health Consensus Conference - Diagnostic Criteria**

Cafe’s-au-lait spots (six or more larger than 5 mm in greatest dimension in prepubertal individuals and larger than 15 mm in postpubertal individuals) Two or more neurofibromas of any type or one plexiform neurofibroma Freckling in the axillary or inguinal region Optic glioma Two or more Lisch nodules (hamartomas of the iris) A distinctive osseous lesion such as sphenoid wing dysplasia or thinning of long bone cortex with or without pseudoarthrosis (5) A first-degree relative (parent, sibling, or offspring) with neurofibromatosis-1 by the above criteria

**Treatment:**

1. Surgery – Subtotal excision of lesion to treat the cosmetic deformity. Difficult to dissect, Cord-like, Woven (6) into the fabric of the tissue. It tends to bleed significantly during surgery. Recurrence of the neurofibromas is common. 2. Chemotherapy (1) 13-cis retinoic acid, alfa interferons & radiotherapy are adjuvants also under evaluation in malignant plexiform neurofibromas

**Conclusion:**

Early surgical intervention of small superficial neurofibroma is uncomplicated. Subtotal resection prevents later disfigurement and functional deficits. Complete resection is difficult. Malignancy is a definite risk.

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**References:**


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