Disfiguring Plexiform Neurofibromatosis

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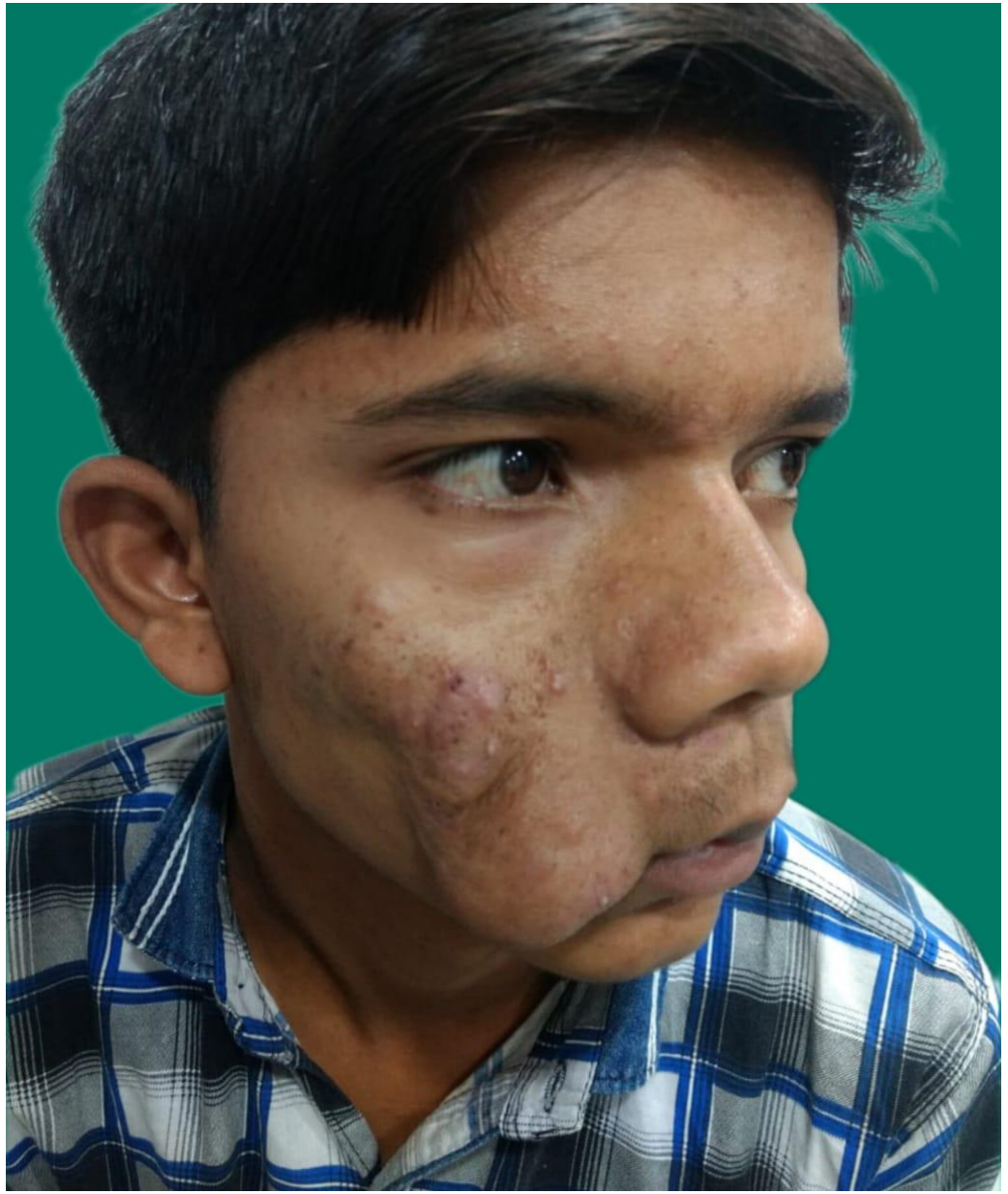
Pre operative



Histopathological Report

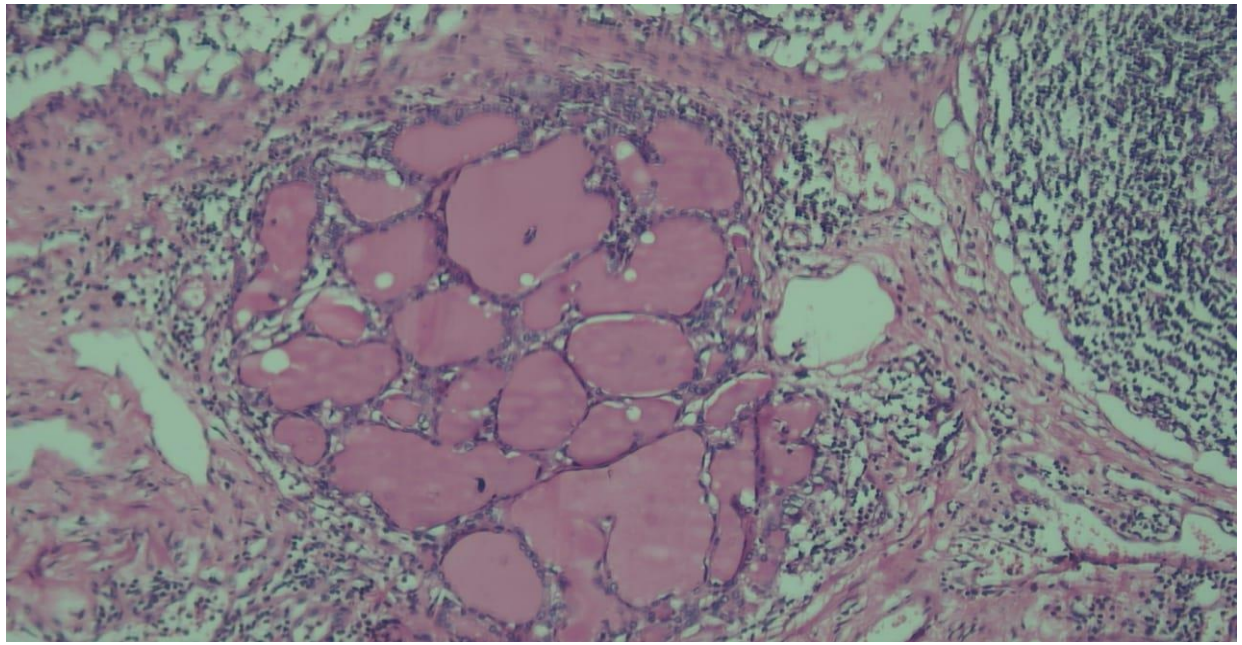
B/2198/19
23/5/19

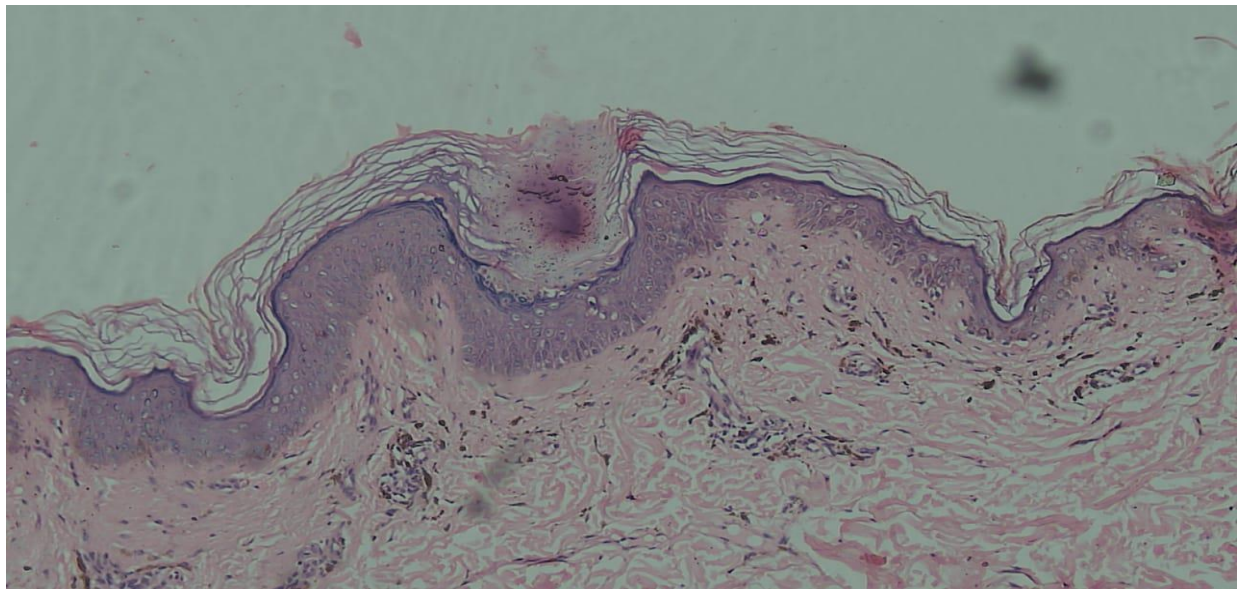
S/B Dr Harsh Kumar (HOD)

Section shows lining stratified squamous epithelium. Underneath stroma shows proliferation of peripheral nerves, fibroblast and collagen. Perineural cells are arranged in plexiform type. At places showing infiltration to muscle tissue. Stroma is also showing mild to moderate mixed inflammatory infiltrate along with many giant cells. No verocay bodies. No palisading of nuclei or hyalinization of vessels

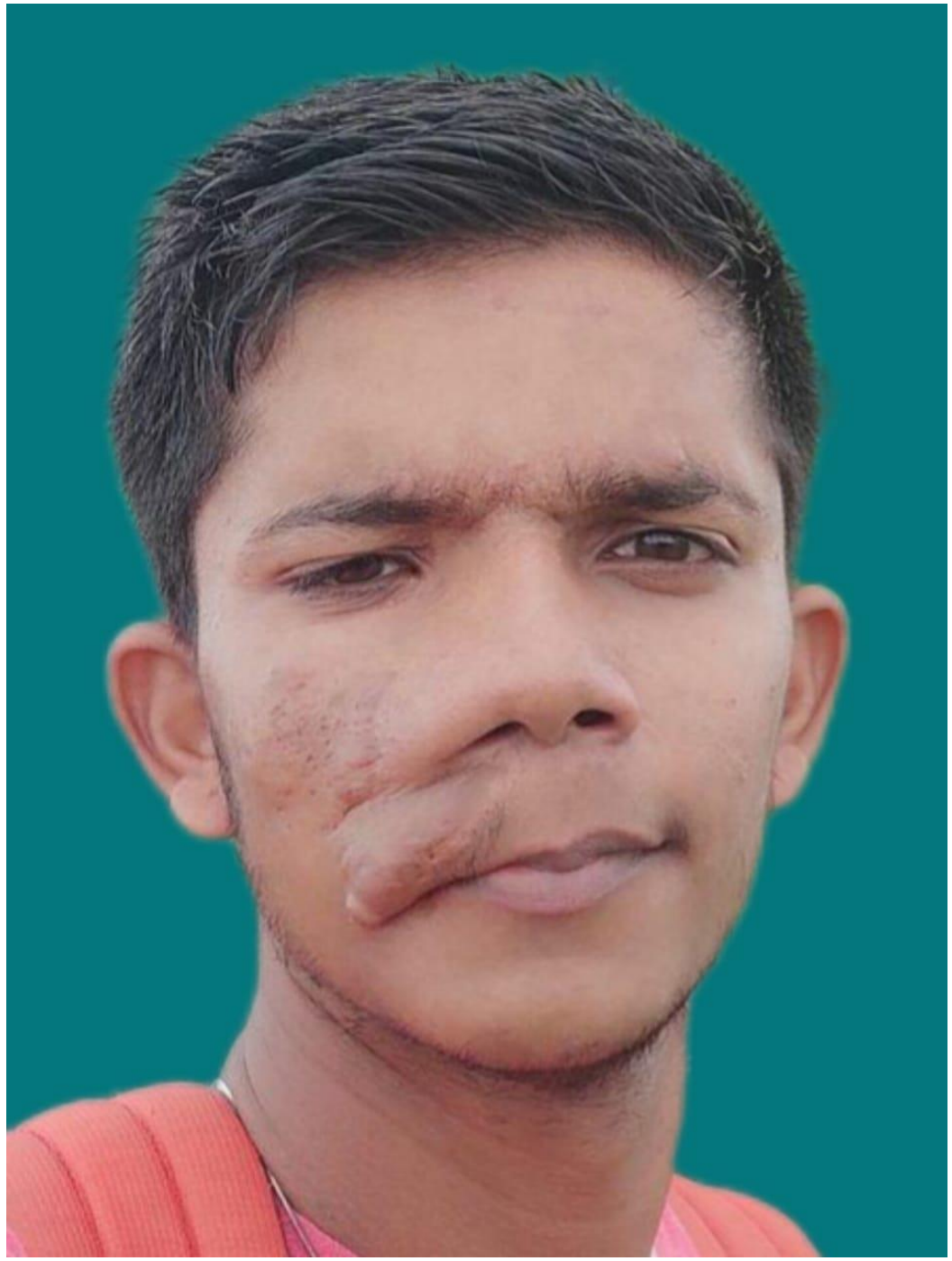
Impression - Plexiform neurofibroma with giant cell reaction.

DR. KOMAL D. SAWAIMUL
Associate Professor
Dept. of Pathology



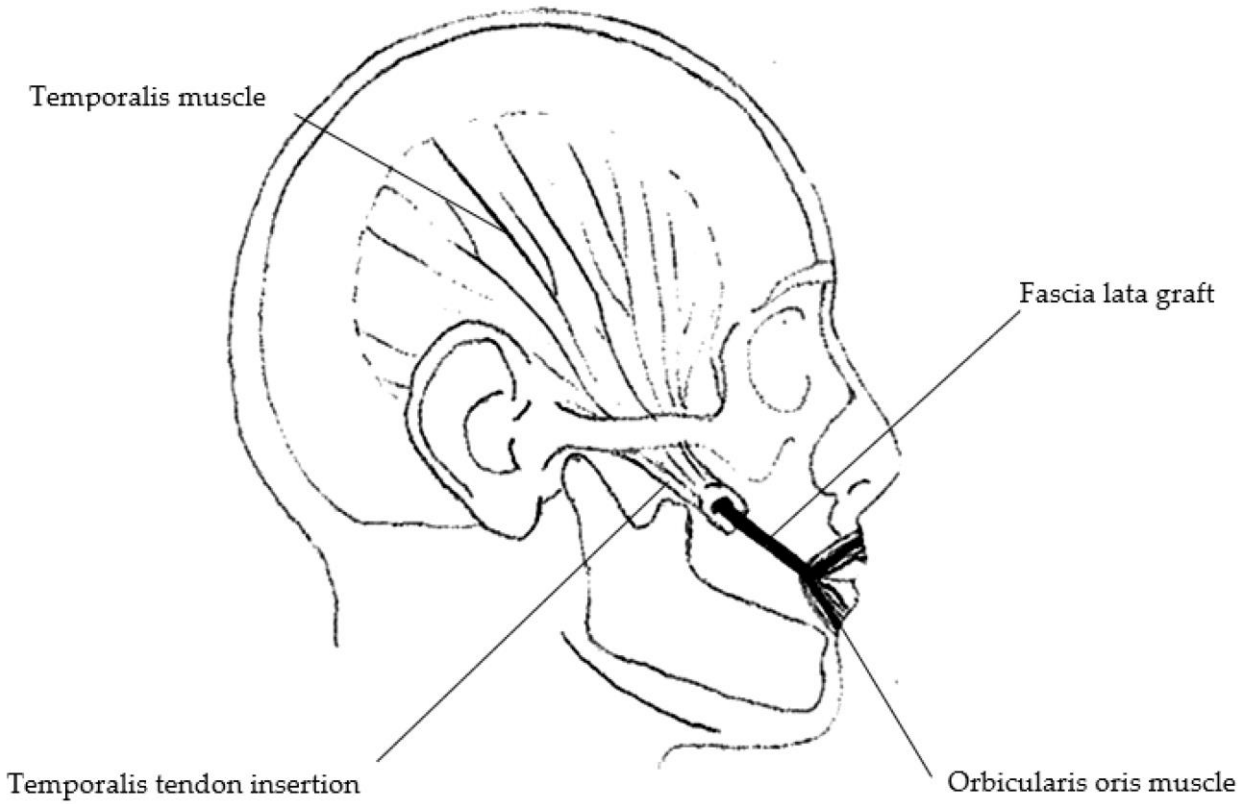


Post 1st Debulking surgery

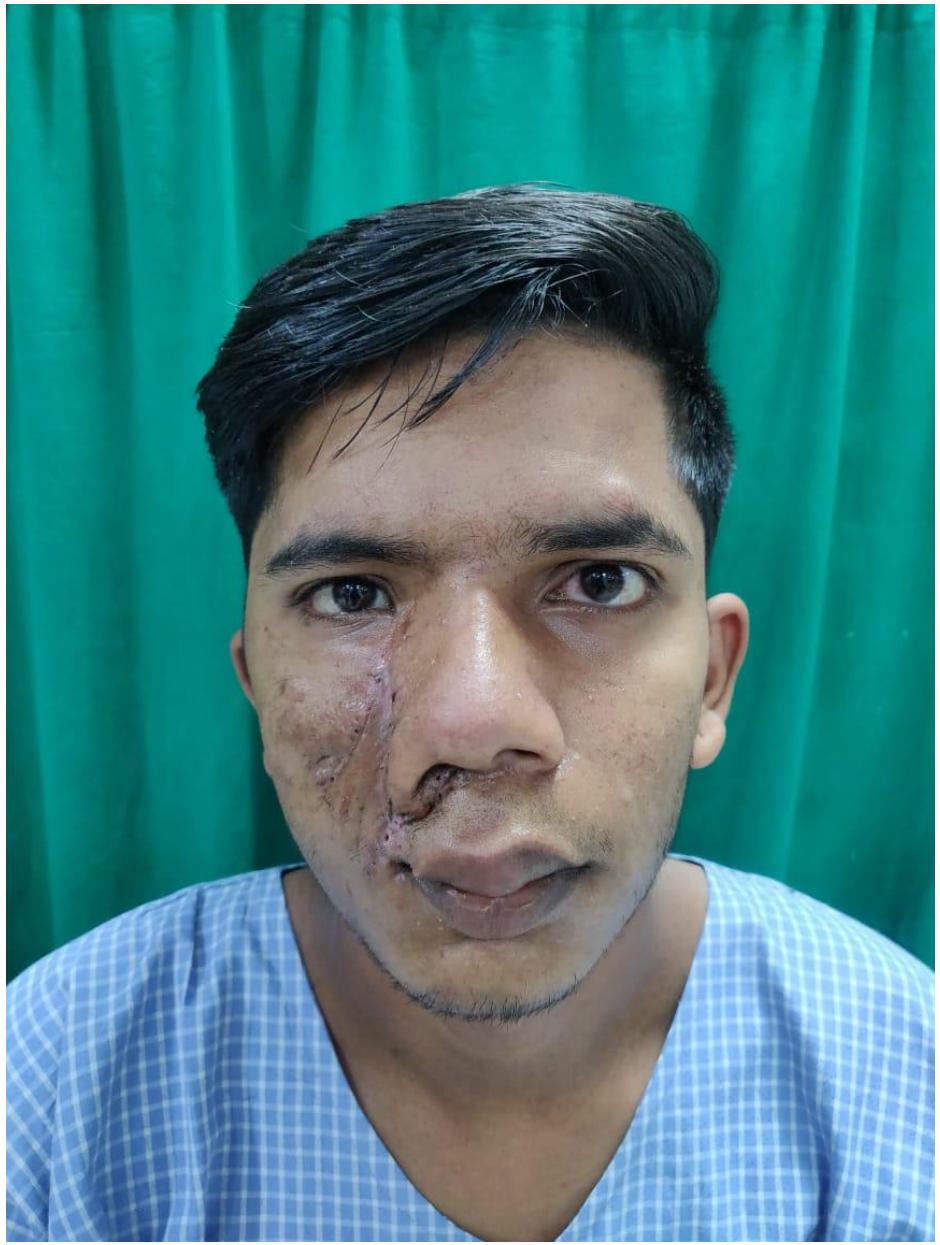


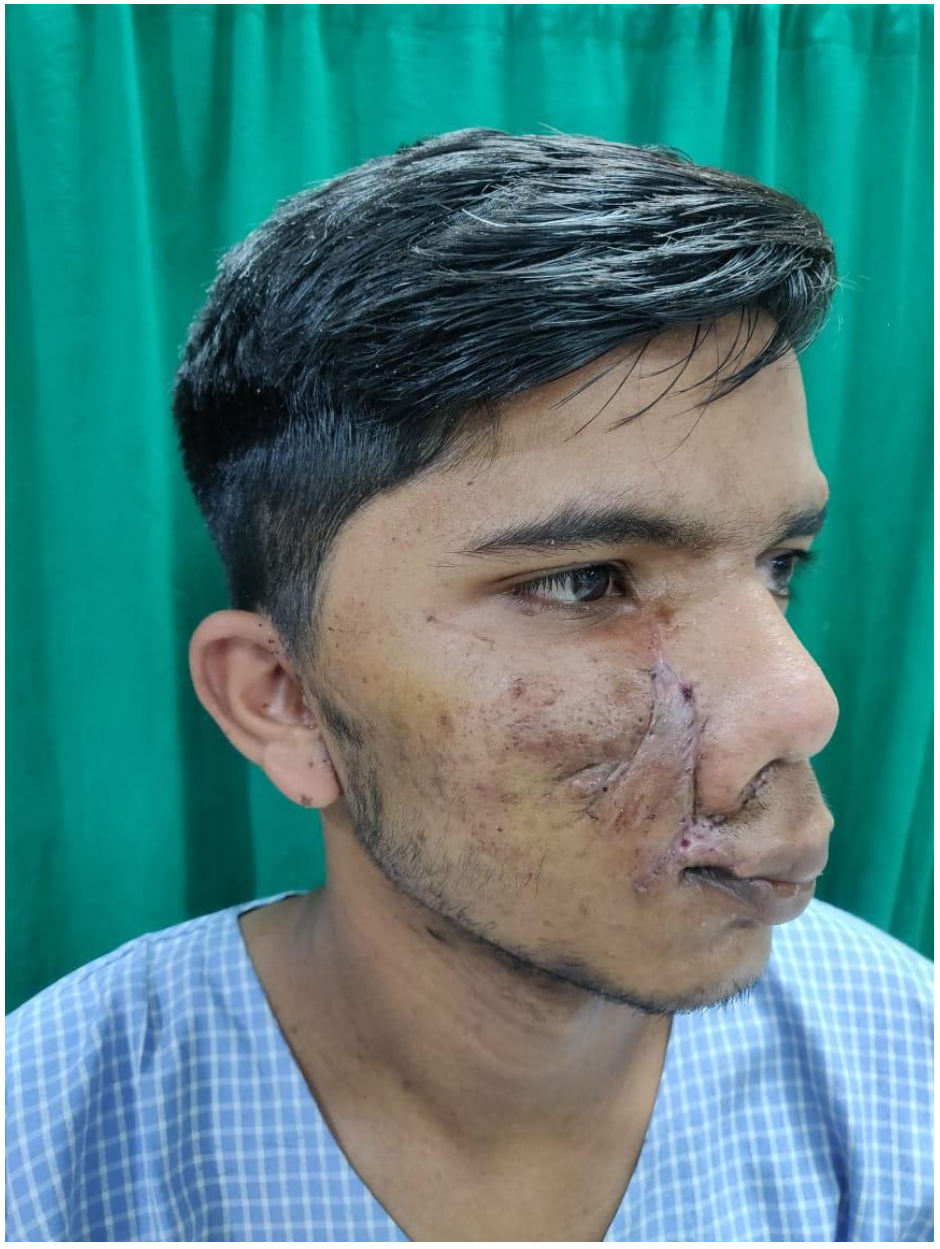
Post 2nd Debulking Surgery

Orthodromic Temporalis Tendon Transfer



Post Lip Repositioning





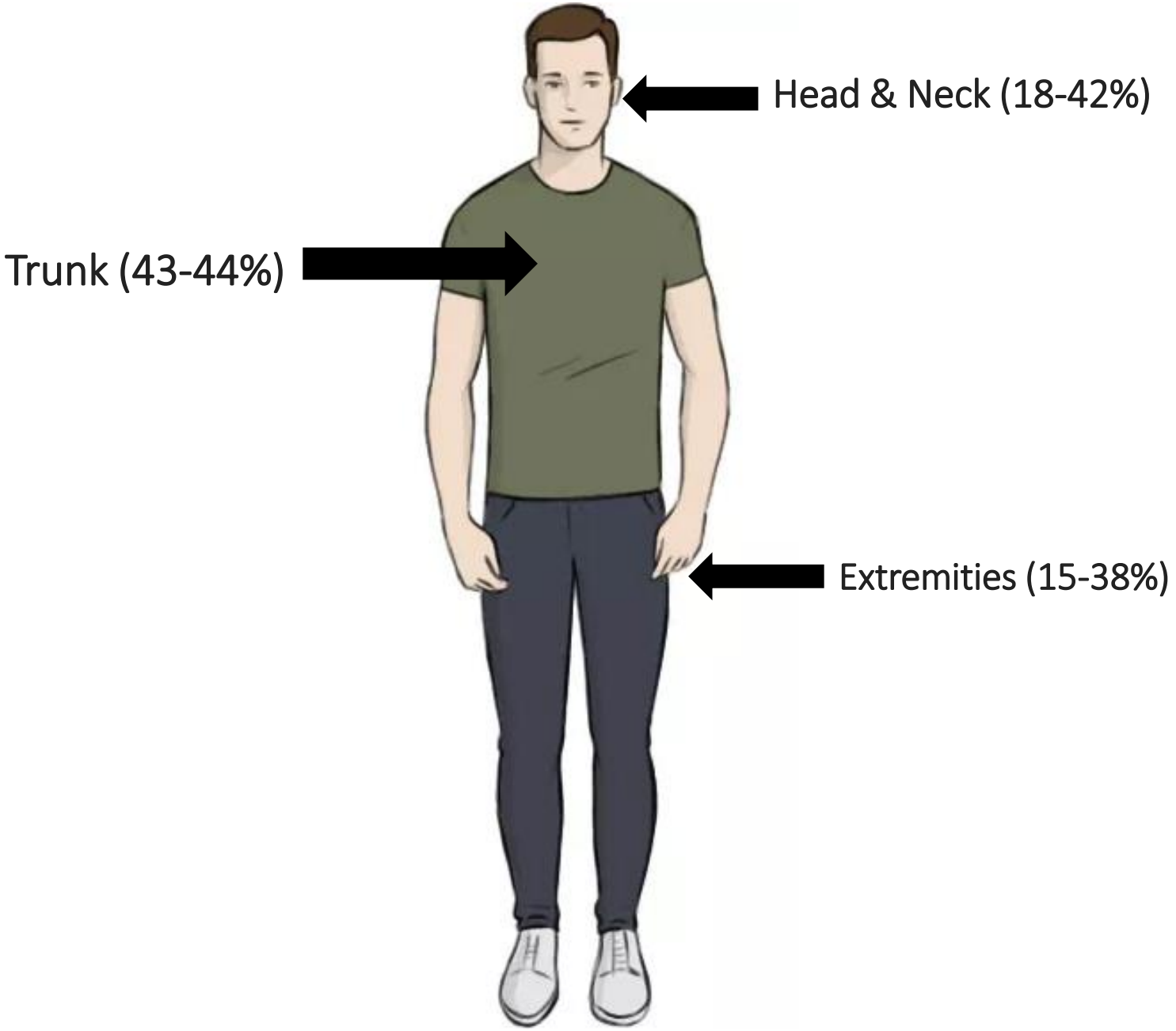
Aim:

Aim of presentation of this case was to highlight the deformities which can be cause by this uncommon condition and how plastic surgery can restore the aesthetic appearance as well as image of confidence.

Plexiform Neurofibromatosis

- Irregular
- Thick
- Non circumscribed tumor
- Peripheral nerve sheath
- Multiple nerve fascicles
- Composed of nerve sheath cells
- Associated with
  - hypertrophy of the overlying soft tissue
  - hyperpigmentation
  - hypertrichosis of the overlying skin

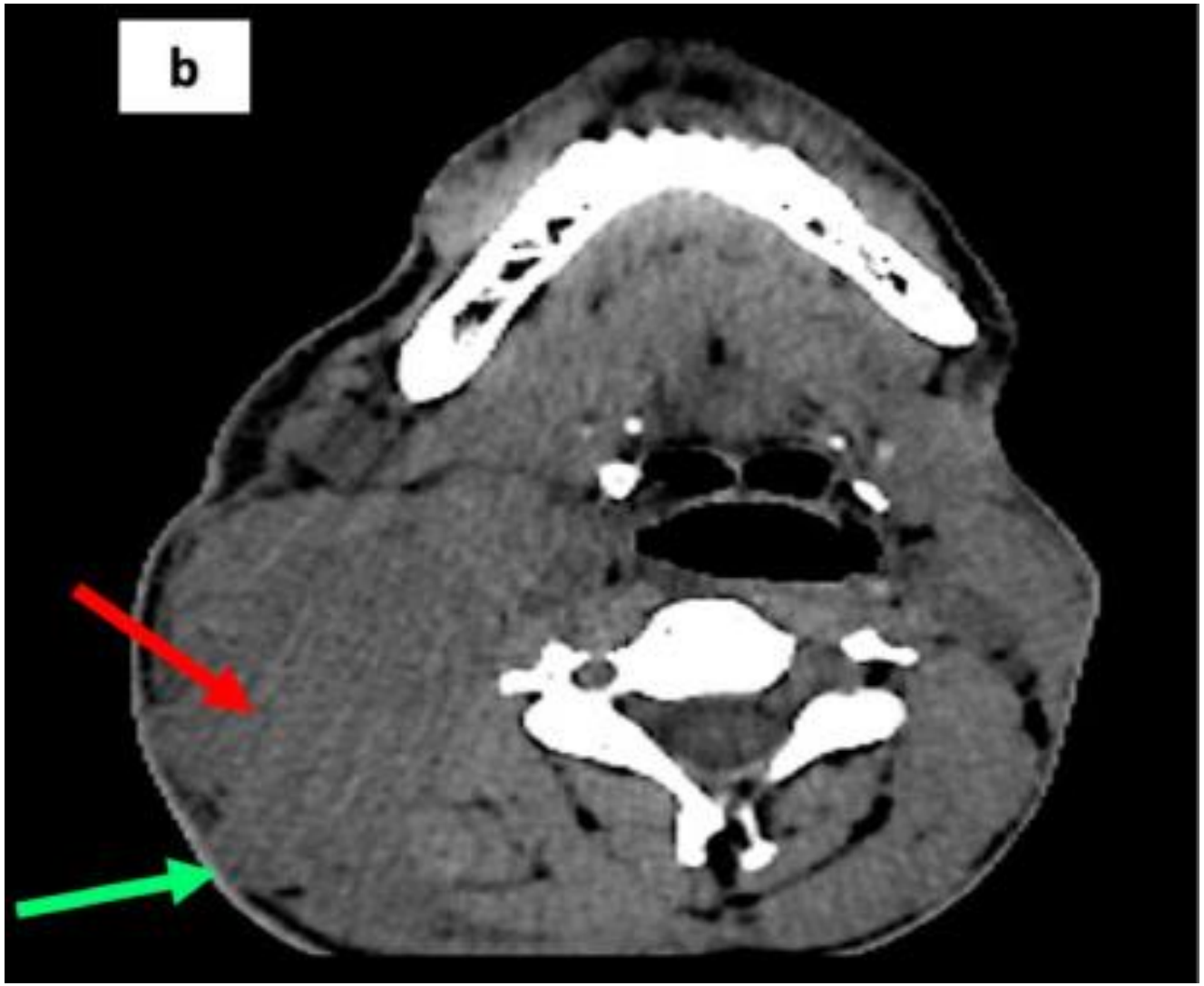
- Incidence
- Characteristics
- Site



Imaging

CT Scan

- low attenuation
  - myelin-lipid content
  - fat entrapment
  - high-water content in endoneurial myxoid tissue

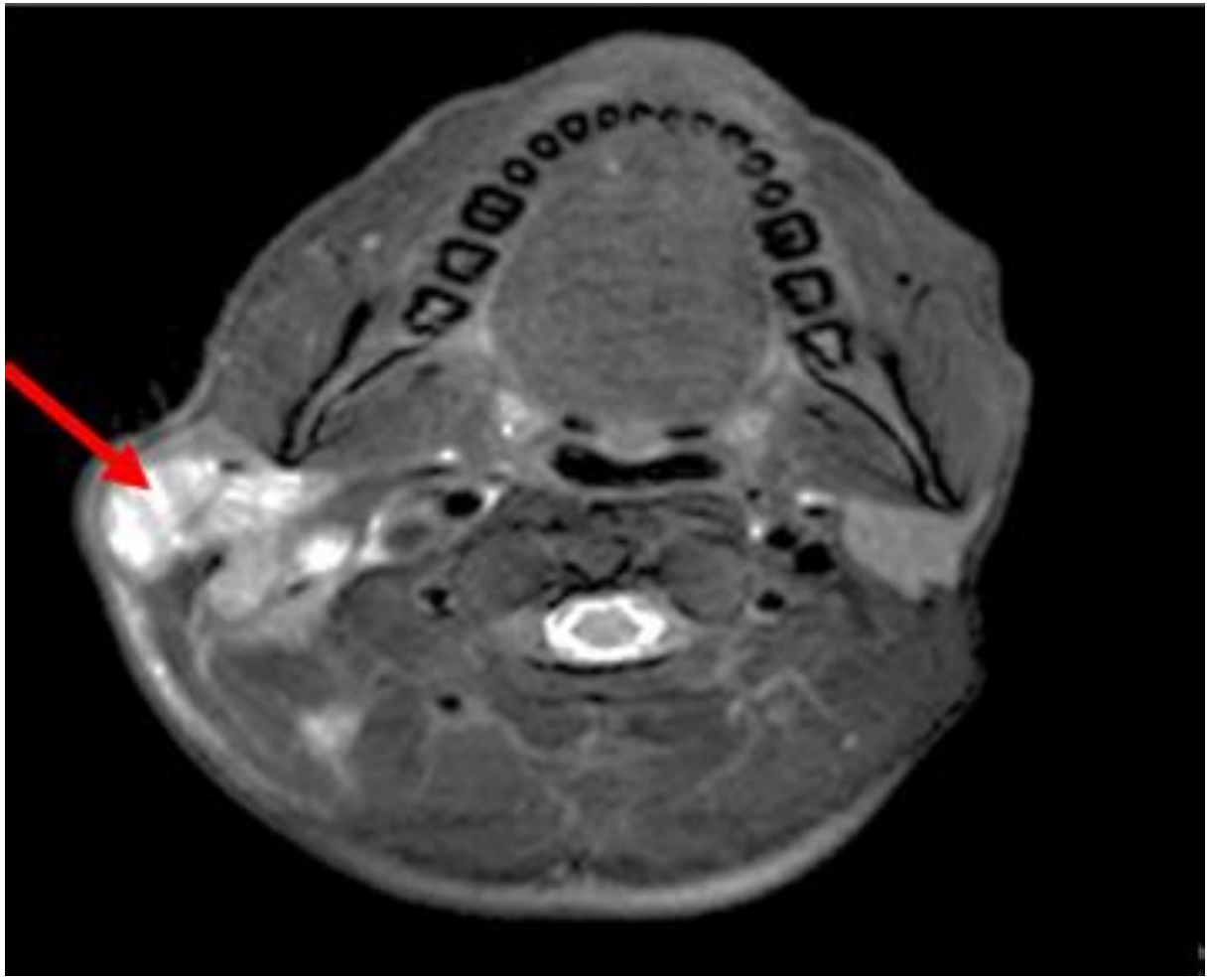


MRI

- lobulated with a hyperintense signal on T2W imaging

CT Angiography

Preoperative Embolisation

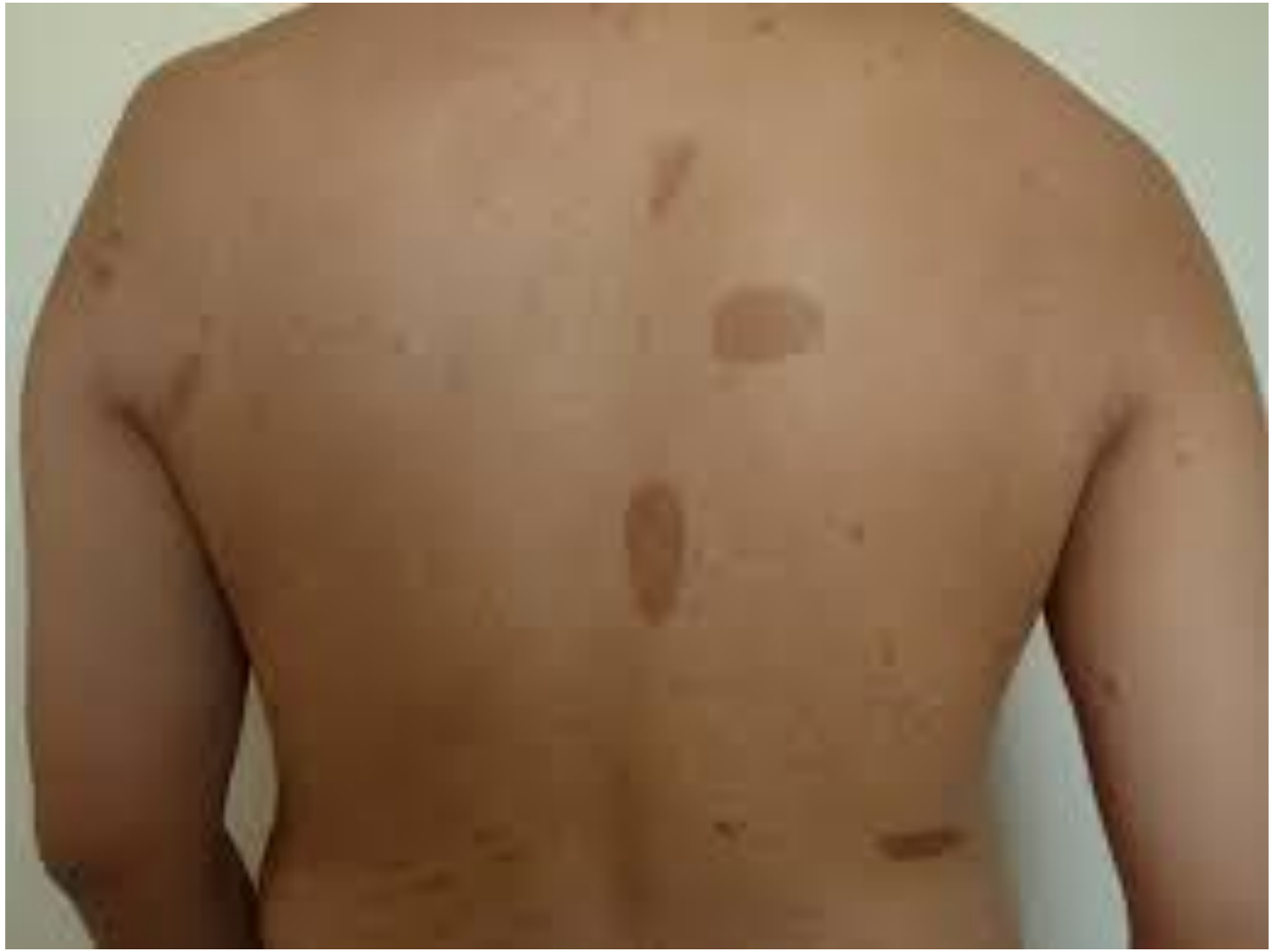


Diagnostic Criteria

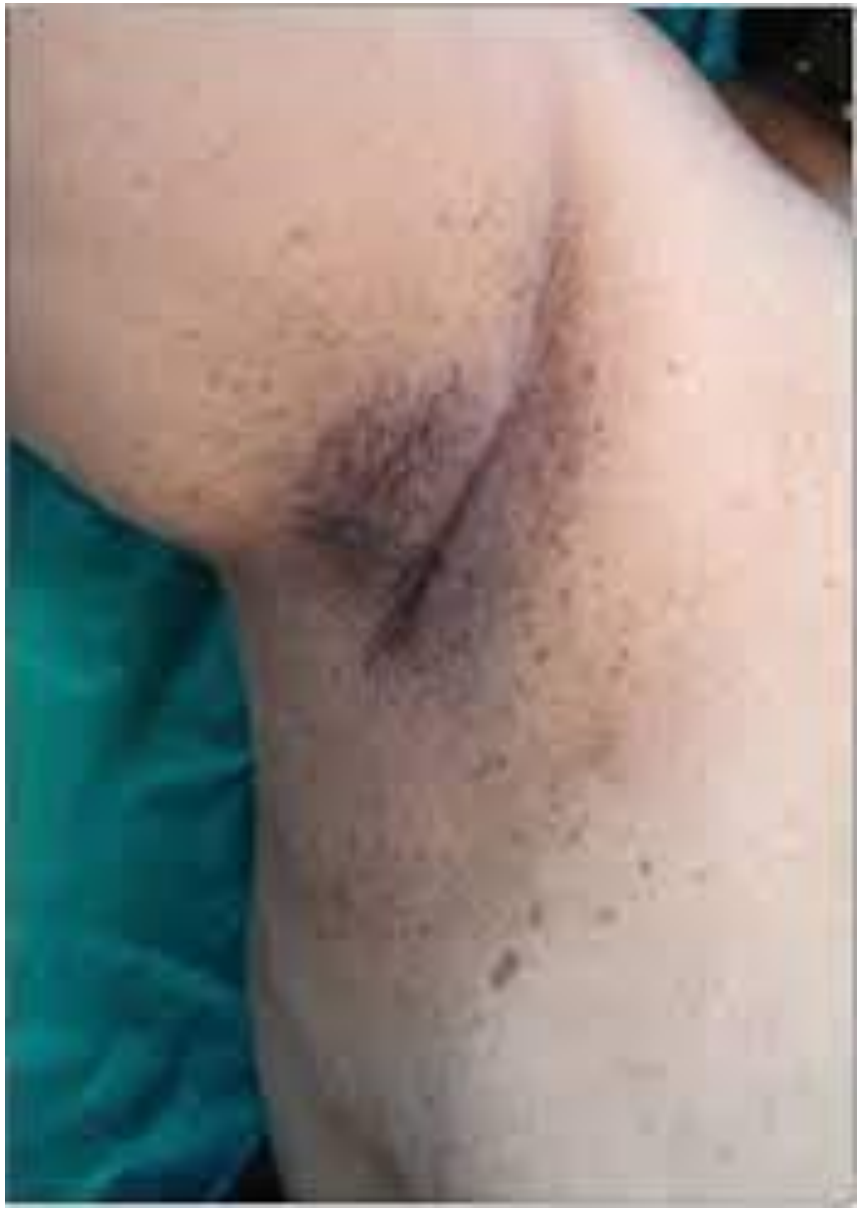
(a) Family History

(b) Café-au-lait macules:

- > 1.5 cm after puberty and
- >0.5 cm in prepubertal individuals



(c) Freckling in the axillary or inguinal region (Crowe sign)

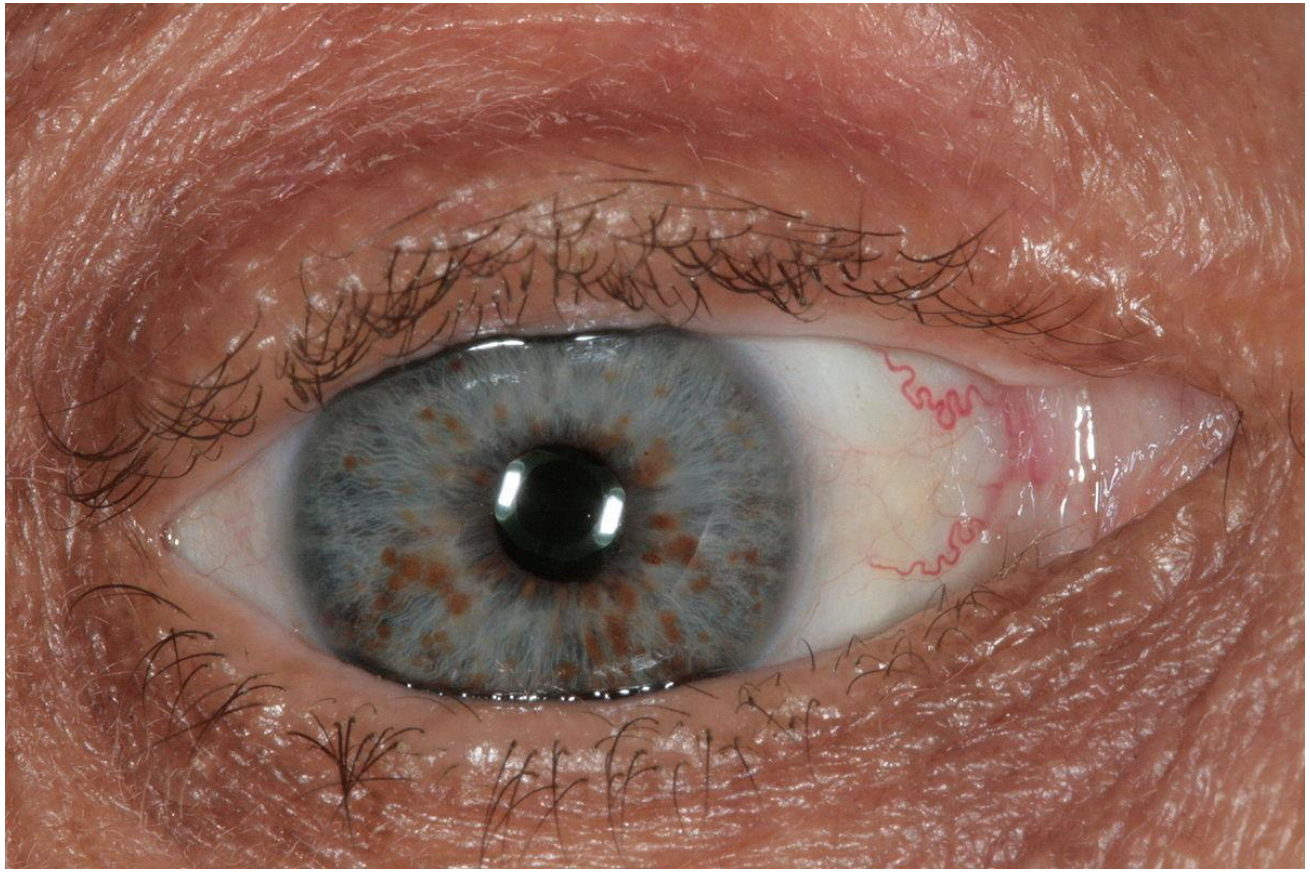


(d) At least 2 neurofibromas of any type or at least 1 plexiform neurofibroma

(e) Optic pathway glioma

(f) At least 2 Lisch nodules (iris hamartoma)

(g) A characteristic bone lesion, which can be: pseudoarthrosis, sphenoid wing dysplasia and thinning of long bone cortex



Difficulties in Surgery

1. Bleeding
2. Nerve Damage
3. Neuroma Formation
4. Infection
5. Altered Sensations
6. Scarring
7. Chronic Pain
8. Recurrence

Thank you