

CASE 2

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A 24 Day old Male child
presented to the Clinic with
Parents Complaining of
Large Left Eye Since Birth.

Examination under anesthesia :

Left side of face showed skin discolouration (hyper pigmentation)

Left eye large and proptosed

Left eye lid thickened

Left cornea large (13mm) but clear

AC - Well formed

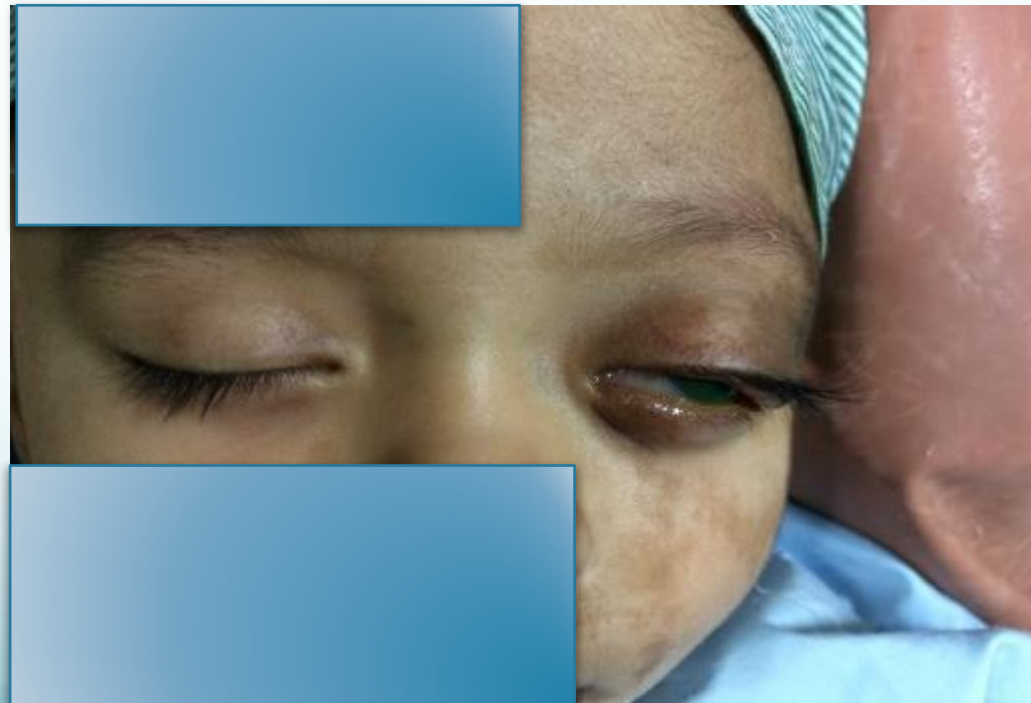
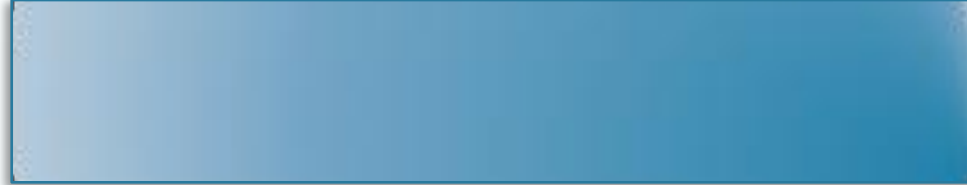
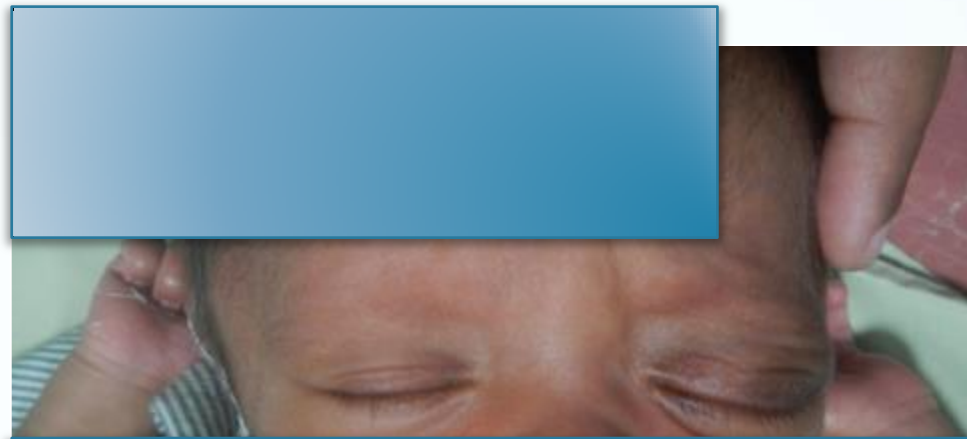
Lens - clear

Pupil - normal

Fundus - disc normal

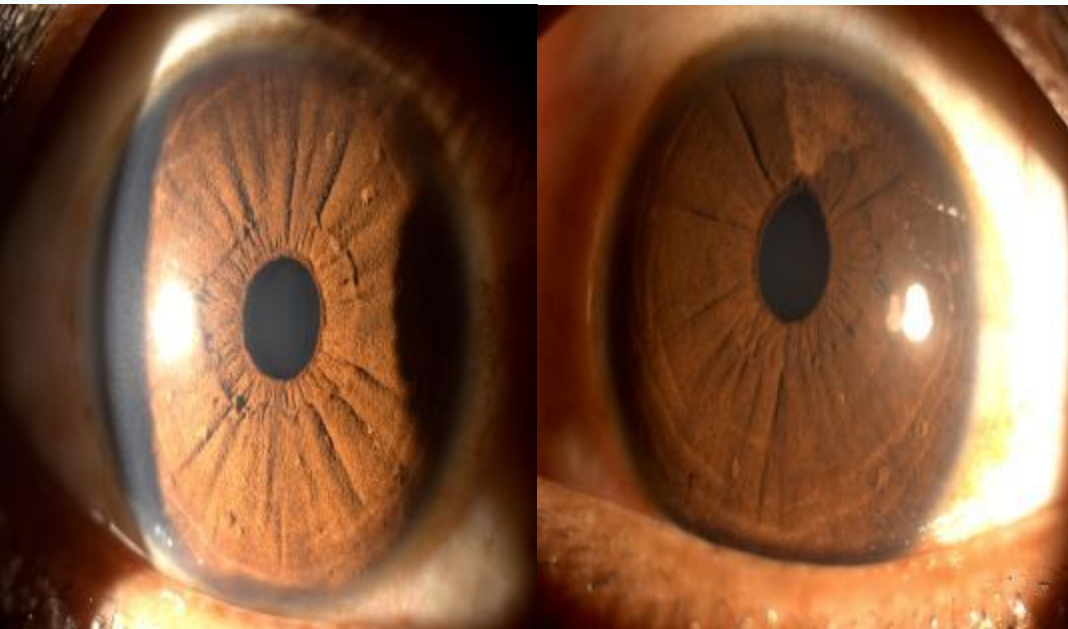
IOP - 12 mm Hg

Right Eye - within normal limits



Family history

- Father showed skin lesion
- Paternal grandmother had nodules on the face and limbs.
- Both had iris nodules and hypo pigmented patches



- Systemic examination of the child revealed similar skin lesions seen on left wrist and right thigh.
- Advice : MRI brain and orbits.
- Provisional diagnosis : Congenital Neurofibroma of left eye.

MRI REPORT

- Enhancing lesion involving left Meckel's cave extending along the juxtaseellar or cavernous sinus region along the superior orbital fissure into the intraconal compartment of left eye.
- Primary lesion is along the left maxillary nerve. The entire lesion measures 22x13x10mm.
- Possibility of Neurofibroma.

The Patient was Advised
a Neurosurgeon's opinion

PATIENT RETURNED AFTER 8 MONTHS!!

Now the parent's complaint was
decreased vision in the left eye.

Nine months since first visit

Examination of left eye

- Cornea ; 16mm in diameter, mild haze (edema).
- AC - normal
- Lens - clear
- Fundus – disc c;d 0.6
- IOP - 30 mmHg
- Right eye - normal



Treatment : Topical - Beta blocker twice daily.

Systemic - Oral acetazolamide.

One month later : IOP controlled, corneal edema decreased.

No Neuro-surgical intervention performed yet.

WHAT WOULD YOU DO NEXT?

What should be the approach for control of IOP?

1) Medical

2) Surgical

[If your answer is option 2 (Surgical), then & only then, please answer sub questions a) & b)]

a) What procedure would you use if your approach was Surgical?

i) Trabeculotomy with Trabeculectomy

ii) Use of a Valve

b) Timing of Surgical Intervention

i) Immediate

ii) After Medical Management Failure

iii) After Neurosurgical Intervention

Thank you !!