

CASE 2

DR MONICA SAMANT
LAXMI EYE INSTITUTE
MUMBAI (INDIA)

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

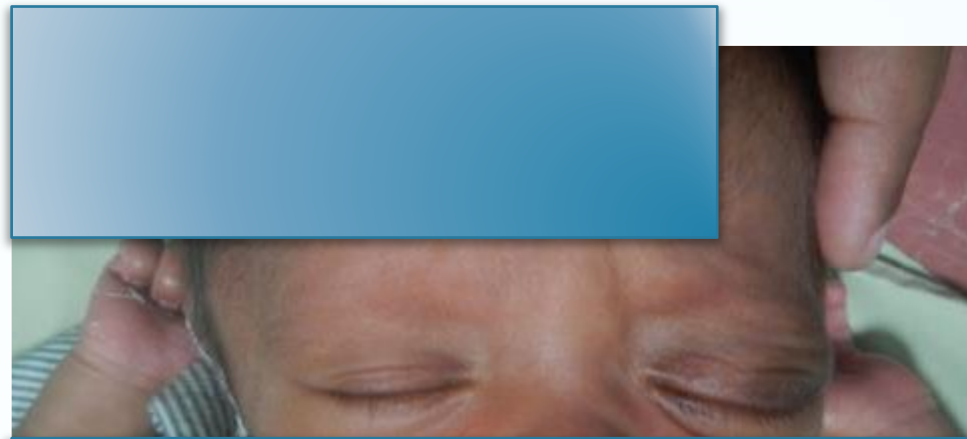
Lens - clear

Pupil - normal

Fundus - disc normal

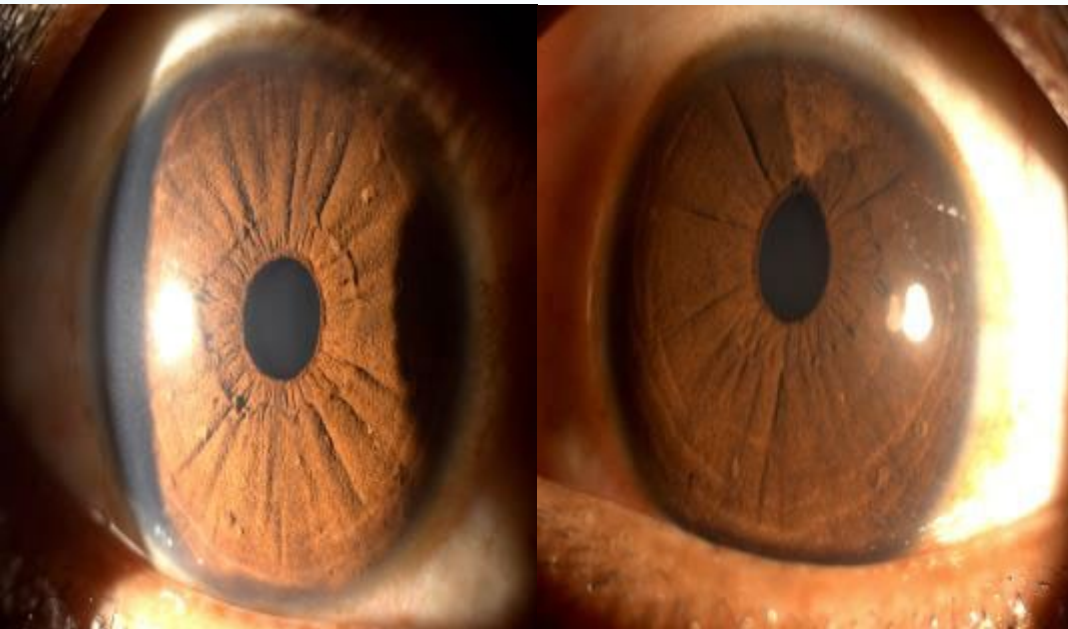
IOP - 12 mm Hg

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 according to you should be the approach for control of IOP?

1) Medical

2) Surgical

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 (if required)

i) Immediate

ii) After medical management failure

iii) After neurosurgery Intervention (if any)

Would you do anything else / differently for such a case?

Could our experts please answer these 2 questions in addition to the answering the questions on the Previous slide?

How would you treat the Proptosis?

What according to you is the role of genetic studies in such cases?

Thank you !!