University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 **2020, Vol. 6(8)**

A CASE REORT OF BROWNS SYNDROME

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Abstract : Browns syndrome is a restrictive myopathy involving the tendon of superior oblique muscle due to abnormalities in the tendon trochlea complex. It may be either congenital or acquired. Here is a case report of a eleven year old girl child who presented with complaints of double vision on upgaze for six months. She had normal anterior and posterior segment findings except for restriction of few extra ocular movements in the left eye with a characteristic chin lift. She was diagnosed to be a case of Browns syndrome on the basis of clinical features and investigations. She was treated with prisms that corrected her diplopia and abnormal head posture.

Keyword : Browns syndrome, Restrictive myopathy, Incomitant strabismus, Hypertropia

CASE REPORT:

PROFILE PICTURE

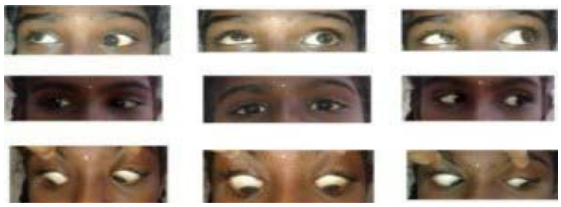
Kousalya, 11 year old female child, came with complaints of double vision since 6 months which was more on looking up with both the eyes. There was no history of trauma, fever, headache, facial pain or joint pain. Her antenatal, perinatal and post natal history was uneventful. General and systemic examinations were normal. On ocular examination, the girl had an abnormal head posture with a chin lift. Eyes did not appear orthophoric. There was right sided hypertropia. Anterior segment findings were normal in both the eyes. Fundus was also normal. The only significant clinical finding was restriction of elevation in adduction, elevation in abduction and elevation of left eye.

ANTERIOR SEGMENT:

RIGHT EYE		LEFT EYE
6/6	VISION	6/6
16	TENSION (mm hg)by NCT	16
Normal	LIDS	Normal
Clear	CONJUNCTIVA	Clear
Clear	CORNEA	Clear
Normal depth	AC	Normal depth
Colour pattern normal	IRIS	Colour pattern normal
Reacting to light	PUPIL	Reacting to light
Clear	LENS	Clear

FUNDUS: Both Eyes Normal

EXTRA OCULAR MOVEMENTS:
RE: Full
LE:



EXTRA OCULAR MOVEMENTS

- Restriction of elevation in adduction
- Mild restriction of elevation in abduction
- Mild restriction of elevation in upgaze

FIELDS: Both Eyes Normal
COLOUR VISION: Both Eyes Normal
ORTHOPTICS:
Cover test :
R / L HYaPERTROPIA L HYPOTROPIA 10 ° Prism Bar Cover Test :
FR : R/L Hypertropia 10
FL : R/L Hypertropia 30

Secondary deviation was more than primary deviation (Incomitant squint) Binocular Single Vision: Normal
DIPLOPIA CHARTING:
Maximum separation of uncrossed L/R images in levoelevation and crossed L/R images in dextroelevation was noted.
MRI ORBIT: Normal
DIFFERENTIAL DIAGNOSIS:
Brown's syndrome Double elevator palsy
FORCED DUCTION TEST:
Test was positive on ELEVATION IN ADDUCTION AND ELEVATION compared to ELEVATION IN ABDUCTION showing it to be due to a restrictive pathology.
FINAL DIAGNOSIS:
Left eye Moderate BROWN'S SYNDROME
MANAGEMENT:

CORRECTION WITH PRISMS
The child was conservatively managed with prisms.
LE : with 4.00 PD base up Prism , head posture became normal with no diplopia.

DISCUSSION:
Brown's syndrome accounts for 1 of every 450 cases of congenital strabismus. The characteristic clinical feature is restriction of elevation in adduction. It was originally thought to be caused by shortening of the sheath of the superior oblique tendon. It is now attributed to various abnormalities of the tendon-trochlea complex like impaired slippage of tendon through the trochlea and developmental anomalies of trochlea.

ETIOLOGY:
Congenital
1. Idiopathic.
2. Congenital click syndrome associated with audible click (due to impaired slippage of tendon through the trochlea) It is unilateral in 90% of cases.

Acquired
1. Trauma to the orbit involving the region of the trochlea (canine tooth syndrome of knapp) or after sinus surgery
2. Tenosynovitis of superior oblique trochlear apparatus
3. Inflammatory as in sinusitis and juvenile rheumatoid arthritis (intermittent Browns) d/t rheumatoid nodules on superior oblique tendon

CLINICAL FEATURES:
1. Deficient elevation in adduction that improves in abduction but often not completely
2. Attempts at straight-ahead elevation usually cause divergence (V pattern).
3. Forced ductions show severe mechanical restriction on attempts to elevate the adducted eye; no/less limitations of elevation in abduction
4. Downshoot in adduction
5. Widening of palpebral fissure on adduction
6. Hypotropia in primary position
7. Compensatory head posture (chin up)

GRADING:

GRADES	ELEVATION RESTRICTION IN ADDUCTION	HYPOTROPIA IN PRIMARY GAZE	DOWNSHOOT IN ADDUCTION
MILD	Present	Absent	Absent
MODERATE	Present	Present	Absent
SEVERE	Present	Present	Present

DIFFERENTIAL DIAGNOSIS:
Paralysis of inferior oblique muscle
DIFFERENTIATING FEATURES:
- "A" pattern in upgaze because of loss of the abduction of the paralyzed muscle in upward gaze
- Over action of superior oblique muscle is present
- Forced duction test : Negative in inferior oblique paralysis

Other differential diagnoses include:
- Monocular Elevation Deficiency (double elevator palsy) Due to paresis or paralysis of ipsilateral superior rectus and inferior oblique muscle. Forced duction testing is useful to differentiate these disorders
- Graves ophthalmopathy
- Congenital fibrosis of inferior rectus muscle.

MANAGEMENT:
CONSERVATIVE :
- Prisms (base up prisms) in small degree deviations.
- Oral /systemic steroids in case of rheumatoid disease.

SURGICAL: SUPERIOR OBLIQUE WEAKENING PROCEDURES:
- SO tendon tenotomy
- SO tendon tenectomy
- SO tendon chicken suture (mercilene, non absorbable)
- SO tendon weakening with silicone expander which is considered to be best because the incidence of superior oblique palsy following surgery is only 10 %.

INDICATIONS FOR SURGERY:
- Diplopia in primary position
- Marked abnormal head posture
- Absence of BSV

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