Abstract:
Cervico-Oculo-acoustic syndrome was first described by Wildervanck in 1952. We report a rare case of Wildervanck Syndrome exhibiting Klippel-Feil anomaly with fused cervical vertebrae, Duane retraction syndrome and conductive deafness (c.f. sensori neural loss).

Keyword:
Wildervanck syndrome, Duane retraction syndrome, Klippel-feil anomaly.

Introduction:
Wildervanck syndrome is a systemic association of Duane syndrome in which a spectrum of motility disturbances occur characterized by retraction of the globe in actual (or) attempted adduction. Limitation of horizontal eye movements in both directions and upshoot (or) downshoot of the affected eye in attempted adduction can occur. Associated features of sensorineural hearing loss and fusion of cervical vertebra can occur as described by Wildervanck.

Case Report:
A 27 years old gentleman presented to our outpatient department with complaints of limitation of outward movement of right eye since childhood. There was no history of defective vision (or) trauma (or) diplopia. He was born out of a second degree consanguineous marriage, delivered normally at full time from an uncomplicated pregnancy. Developmental milestones were normal. There was no history of similar features in any of the family members. Clinical examination revealed limitation of abduction of right eye, retraction of the eyeball into the socket on adduction with associated palpebral fissure narrowing, elevation (or) upshoot of the right eye on adduction (leash phenomenon) and poor convergence. Both eyes best corrected visual acuity was 6/6. Both eyes anterior segment and fundus examination was normal.
Vertebral segmentation occurs between the fourth and eighth weeks of gestation and any impairment in differentiation of the mesoderm may relate not only to cervical abnormalities but also to the combination of rare cardio vascular abnormalities. (6) Indications for surgical intervention in Duane syndrome includes primary position deviation, abnormal head posture, marked globe retraction and large up or downshoots. Mostly patients are treated conservatively. Although most patients of Duane retraction syndrome have only ocular features, many associate systemic defects have been observed, including Goldenhar syndrome, Wildervanck syndrome, Okihiro syndrome, Holt-oram syndrome and Morning glory syndrome. So it becomes important to perform a thorough systemic examination to rule out other associated congenital anomalies.

References
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