Abstract: OBJECTIVE: We report a rare presentation of bilateral vestibular schwannomas with left eye ptosis with ophthalmoplegia. CASE REPORT: An 18 year old female patient was referred from Ophthalmology department to ENT department with one year history of left eye lid drooping and hard of hearing left ear. Pure tone audiogram showed left ear moderate SN hearing loss and right ear mild SN hearing loss. Brain Stem Evoked Response Audiogram showed wave III and wave V latency on both sides more on left side. Gadolinium enhanced MRI brain showed bilateral acoustic neuroma with neurofibromatous changes in the left eye retro orbital area with medial rectus hypertrophy. CONCLUSION: Ptosis with ophthalmoplegia has rarely been reported in NF2. In our case described, left eye ptosis with ophthalmoplegia was caused by mechanical effect of mass in retro orbital area. In this case since the tumors on both sides more than 1.5cm, the best option is regular follow up eventhough she had moderate hearing loss on the left side. Patient was advised for correction of left eye ptosis, but she was not concerned about it and deferred surgery. So she was advised regular follow up annually for clinical, audiological and radiological evaluation.

Keyword: Bilateral Vestibular Schwannomas, Brain Stem Evoked Response Audiogram, Gadolium Enhanced MRI

INTRODUCTION: Patients with NF2 typically present with either unilateral or bilateral hearing loss, with or without balance problems. NF2 is a disorder usually diagnosed later in life since the features are subtle in children. The hallmark is bilateral vestibular schwannomas, which may not appear until after the second decade. Bilateral vestibular schwannomas with left eye ptosis with ophthalmoplegia is a rare case and only a few cases reported in literature. Half of the affected individuals inherited the disorder from an affected parent and half seem to have a mutation for the first time in their family.

CASE REPORT: An 18 year old female patient referred from Ophthalmology department to ENT department with one year history of left eye lid drooping and hard of hearing left ear. In Ophthalmology department they did visual acuity testing and fundus examination and found to be normal. She had no relevant past or family history. On examination, patient had left eye lid drooping and ophthalmoplegia(fig.1). Both ears tympanic membrane intact. Tuning fork test on both ears Rinne positive, Weber lateralized to right ear and ABC reduced on both ears. Both facial nerves intact. Left eye showed reduced corneal sensation with jerk nystagmus. Both eyes pupillary reflex present. Other cranial nerves found to be inact. Sensory system found to be intact, except absent corneal reflex left eye. Motor system intact. Examination of her nose, neck, oropharynx was unremarkable.

Fig.1 Patient photograph showing left eyelid ptosis

Pure tone audiometry showed left ear moderate SN hearing loss and right ear mild SN hearing loss. We did BERA and found wave III and wave V latency on both sides more on left side suggestive of CP angle tumor (fig.2 and3). Patient also undergone speech discrimination score (SDS) and caloric test, found to be normal.
**DISCUSSION:**

Neurofibromatosis type 2 is an autosomal dominant disorder where hallmark is bilateral vestibular schwannomas, which is related to deletion defects from chromosome 22. Epidemiologic studies place the incidence of NF2 between 1 in 40,000 live births and 1 in 87,410 live births. The NIH Consensus Committee has defined clinical criteria for NF2; namely (i) bilateral eight nerve masses or (ii) a first degree relative with neurofibromatosis 2 and either a unilateral eight nerve mass, or two of the following: neurofibroma, meningioma, glioma, schwannoma or juvenile posterior subcapsular lens opacity. The patient reported here fulfills the criteria for the diagnosis of NF2. Patients with NF2 typically present with either unilateral or bilateral hearing loss, and most have evidence of disturbed vestibular functions. NF2 has been associated with multiple central nervous system tumors, the most common of which are intracranial meningiomas, spinal tumors, and optic gliomas (in addition to cataracts). Intracranial schwannomas most frequently affect the eight nerve. The trigeminal nerve is the next most frequently affected. Similarly, involvement of more than one nerve should warrant a risk up for NF2. Cutaneous markers are rare in NF2 as compared to NF1. CNS lesions are seen virtually in all cases include neoplasms (cranial nerves and meninges), non neoplastic intracranial calcifications and spinal cord nerve root tumors. All NF2 patients and their families should have access to genetic testing because presymptomatic diagnosis improves the clinical management of the disease. Some clinical manifestations of NF2, such as ocular abnormalities, can be detected in infancy, therefore, clinical screening for at-risk members of NF2 families can start at birth, with the first MRI brain scan at 10-12 years of age. Minimal interference, maintenance quality of life and conservation of function or auditory rehabilitation are the corner stones of NF2 management. So initial evaluation of individuals with NF2, such as ocular abnormalities, can be detected in infancy, therefore, clinical screening for at-risk members of NF2 families can start at birth, with the first MRI brain scan at 10-12 years of age. Minimal interference, maintenance quality of life and conservation of function or auditory rehabilitation are the corner stones of NF2 management. So initial evaluation of individuals with NF2, such as ocular abnormalities, can be detected in infancy, therefore, clinical screening for at-risk members of NF2 families can start at birth, with the first MRI brain scan at 10-12 years of age. Minimal interference, maintenance quality of life and conservation of function or auditory rehabilitation are the corner stones of NF2 management. So initial evaluation of individuals with NF2, such as ocular abnormalities, can be detected in infancy, therefore, clinical screening for at-risk members of NF2 families can start at birth, with the first MRI brain scan at 10-12 years of age. Minimal interference, maintenance quality of life and conservation of function or auditory rehabilitation are the corner stones of NF2 management. So initial evaluation of individuals with NF2, such as ocular abnormalities, can be detected in infancy, therefore, clinical screening for at-risk members of NF2 families can start at birth, with the first MRI brain scan at 10-12 years of age. Minimal interference, maintenance quality of life and conservation of function or auditory rehabilitation are the corner stones of NF2 management. So initial evaluation of individuals with NF2, such as ocular abnormalities, can be detected in infancy, therefore, clinical screening for at-risk members of NF2 families can start at birth, with the first MRI brain scan at 10-12 years of age. Minimal interference, maintenance quality of life and conservation of function or auditory rehabilitation are the corner stones of NF2 management.
So tumor resection in a hearing ear should be deferred until the development of neurologic symptoms. Tumor resection in a non-hearing ear should be performed as clinically indicated with a focus on preserving other cranial nerve function. Radiation therapy for vestibular schwannomas has become popular in recent years. But the potential complications include facial weakness, trigeminal neuropathy and vestibular dysfunction. In addition, there is increased risk of developing a secondary malignancy after treatment especially with radiotherapy. This patient met the criteria and diagnosed as NF2 even though spinal cord MRI did not reveal any tumors. Patient had hearing loss in the left ear and the size of tumor is > 2cm on the left side. Patient not so concerned about left eye ptosis. However, patient's parents were counselled regarding the nature of the disease and attendant complications. In this case since the tumors on both sides more than 1.5cm, the best option is regular follow-up eventhough she had moderate hearing loss on the left side. Patient was advised for correction of left eye ptosis, but she was not concerned about it and deferred surgery. So she was advised regular follow up annually for clinical, audiological and radiological evaluation.

REFERENCES: