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A Case Report on a 15-Year Old Boy withNeurofibromatosis with Type-2 Post-Operative Excision of Bilateral Vestibular Schwannoma

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Abstract:
Background: Bilateral vestibular Schwanomas, also known as acoustic neuromas, are most commonly associated with the genetic condition neurofibromatosis type 2 (NF2). These are benign (noncancerous) tumours that form on balance and hearing nerves of the inner ear. This rare disorder instance of NF2 is discussed, as well as its Management and consequences.
Presenting complaints & investigations:A 15-year-old child was seen at Tertiary care rural hospital, wardhawith complaints of hearing loss for the past six years and gait instability for the past four months. It's also linked to trouble speaking and eating, dizziness, impaired vision, and skin schwannoma on the hand, foot, belly, and back, which has been unpleasant for three months**.**
The Main diagnosis, therapeutic intervention & outcome: Following a general physical examination and investigation report, a case of Neurofibromatosis type 2 was found, and Management for surgery was done. Under NIM monitoring, left RetromastoidSuboccipital Craniotomy and excision of left vestibular Schwannoma (CP angle Tumour) were performed. Tab. Levera 500mg twice daily, Tab. Risperidone 2 mg, Syp. Tusqx in TDS, Inj. Ceftriaxone, Inj. Neomaol was given 1gn in SOS, protein powder for supplementary was given.
Conclusion: He responded to both medicine and physician's counselling. His recovery was good.

Keywords: Neurofibromatosis type-2, benign tumour, Retromastoid Suboccipital Craniotomy, vestibular Schwannoma.

INTRODUCTION:

Neurofibromatosis type 2 is a condition that causes noncancerous tumours to form in the nervous system. [1]Vestibular schwannoma and acoustic neuromas are the most prevalent tumours linked with neurofibromatosis type 2. These tumours occur along the nerve that transmits data from the inner ear to the brain (the auditory nerve). This syndrome is also associated with tumours that develop on other nerves. It's an autosomal dominant multiple neoplasia syndrome caused by mutations in the NF2 tumour suppressor gene, which can be found on chromosome 22q.[2]
Bilateral Vestibular schwannomas or acoustic neuromas are the most common manifestations of Neurofibromatosis Type 2.[3]The number of people born with a rare condition is estimated to be between 1:25,000 and 1:40,000, with a prevalence of 1 in 60,000. Males and females are both affected in equal amounts. It is transmitted autosomically and is connected to chromosome 22q12.It occurs once in 25 000 live births and has a nearly 100% penetrance rate by age 60. For neurofibromatosis type 2, half of the patients inherit a germline mutation from an affected parent, and the other half acquire a de novo mutation. [4]
On chromosome 22, the genes implicated in the development of the aforementioned disorders are near together. Due to its inadequate penetrance, Schwannomatosis has a lower risk of transfer to children than NF 2.

Patient Information:

A 15-year-old male child was admitted in pediatric ward, with the main complaint of poor hearing for the previous 4-5 years and gait instability for the last 4 to 5 months. It's also been connected to difficulty speaking and eating during the past 5-6 months, as well as balance concerns, foggy eyesight, and sedentary behaviour. Because he was a known case of neurofibromatosis 2, he was brought to ourhospital for additional treatment.

Primary concern & symptom of a patient:
According to the mother, the child was diagnosed with k/c/o neurofibromatosis 2 in 2015 when he was taken to private hospital for an eye examination. On MRI, multiple meningiomas were discovered in the left prepontine cistern, anterior tentorial region, and left anterior cavernous sinus, as well as posterior flattening and larger eye diameter. According to the patient, several swellings all over his body are growing in size, are painful, and are not receding or diminishing.The mother also claims that she has had difficulty walking and speaking for the past 5-6 months due to swelling on the dorsum of his tongue, that he has an impaired hearing for the past 4-5 years, and that he has difficulty eating and speaking for the past 5-6 months due to swelling on the dorsum of her tongue. His vision has been fuzzy for the past six months. Other symptoms included a fever, vomiting, dizziness, weakness, and others. He was come to the tertiary care hospital for more testing and treatment.

Surgical History:A patient with Neurofibromatosis Type 2 underwent surgery, which included the removal of a left Retromastoid Suboccipital Craniotomy and the removal of a left vestibular Schwannoma (CP angle Tumour) under NIM monitoring.

Medical, family & psychological History:
This patient had a history of head trauma. A CT brain scan revealed multiple, high contusions in the lateral frontal lobe, as well as a 3-5mm subdural hematoma in the left parietal region, with no e/o any midline shift, as a result of an RTA six years ago. For subarachnoid haemorrhage and renal tubular acidosis, the patient was hospitalized for ten days and treated conservatively. He is a member of joint family that lives together. His family is made up of six members. The entire family is in excellent health. With both of his parents, he has maintain good relationship.

Physical examination and Clinical Findings:
During the entrance physical examination, the patient was alert, cooperative, and well-oriented to time, place, and person. He had a troubled and depressed expression on his face. With a thin physique made and suitable sanitation, he was afebrile and vitally stable. Her BMI was 18.37kg/m2; she weighed 36 kilograms and stood 1.32 meters tall. On S1 and S2, his bilateral lungs were clean, and his chest and abdomen tests were normal.Multiple elevated soft, movable swellings over the body that are growing in size and are not regressing or disappearing.
According to the mother, the patient was diagnosed with k/c/o neurofibromatosis 2 in 2015 when she was sent to sevagram hospital for an eye evaluation. Acoustic Schwannoma, meningiomas in the left prepontine cistern, in the anterior tentorial area, and the left anterior cavernous sinus, as well as posterior flattening and increased eyeball width, were discovered on MRI. According to the patient, several swellings all over his body are growing in size, are painful, and are not receding or diminishing. The mother also claims that she has had difficulty walking and speaking for the past 5-6 months due to swelling on the dorsum of her tongue, that she has had an impaired hearing for the past 4-5 years, and that she has had difficulty eating and speaking for the past 5-6 months due to swelling on the dorsum of her tongue.

Diagnostic assessment:
Neurofibromatosis type 2 with vestibular Schwannoma was discovered after a physical examination and study. RBC was low 3.60 cu/cu.mm,Total WBC Count 15000cu.mm was increased, and total platelet count was 3.06 lac/cumm in peripheral blood examination. The LFT and KFT were both within normal ranges. **CT-brain plain in a post operated case of** Extracalvarial soft tissue swelling in the right occipito-parietal region, blood density collection in the left cerebellopontine angle with perilesional oedema causing a mass effect in the form of displacement of the midbrain, pons, vermis, superior and middle cerebellar peduncle to the contralateral side, ill-defined hyperdense lesion in the right cerebellopontine angle, and widening of the bilateral internal acous**.USG Abdo-Pelvis** and **2D ECHO** were done s/o normal study. **MRI brain contrast** was done suggestive of heterogeneously enhancing lesions in Bilateralcerebellopontine angle with mass effect as described above most likely to be Bilateralvestibular Schwannoma. **Histopathological Report** Section from given tumour lesion shows histopathological features suggestive of Schwannoma / Neurilemmoma. **Ophthalmologist Opinion** was taken for lisch nodules s/o normal study. **ENT and OMFS opinion** was taken for difficulty in swallowing and speech s/o no active intervention from OMFS side. ENT surgeon advised BERA. **Brain stem-evoked response audiometry** s/o no 5^{th} wave formation at 110 dB in both the ears. **The psychiatrist's opinion** was taken i/v/o behavioural abnormalities who advised Tab. Risperidone was started on the same.

Diagnosis: After a physical examination and investigation doctor diagnoses a case of neurofibromatosis (nonmalignant) type - 2 with post-op excision of bilateral vestibular schwannoma bilateral acoustic schwannoma (left larger than right),
Prognosis: Child's prognosis was satisfactory.NF2 remains a life-limiting and life spoiling condition.

Therapeutic intervention:
The patient was treated with medical and surgical care. Tab. Levera 500 mg twice a day, tab. Amlodipine, Tab. Sodamint and Tab. Diamox., local application of the t-bact ointment, Tab. Risperidone 2 mg, Tab. Clonazepam Syp. Tusqx in TDS, Inj. Ceftriaxone in antibiotic, Inj. neomaol was given 1 gm in SOS, protein powder The patient is now being treated for Neurofibromatosis type 2 (NF2). The child was allowed to eat and was prescribed Tab. levera.Regarding behavioral abnormalities, psychiatrist opinion was done, who prescribed Tab. Risperidone, and the Tab. Clonazepam for delusions.The patient was intubated, kept on NBM, and started on IV fluids, inj. Ceftriaxone, inj. Amikacin, inj.Neomol, inj. emset, inj. levera, inj. methylprednisolone, inj. midaz, inj. Fentanyl drip, and Tab. labetalol because his blood pressure was high. The ventilator was subsequently removed from the patient. The patient was extubated on post-operative day 1 and given inj. vancomycin i/v/o facial swelling in substitution for inj. amikacin. The RT feeds have begun.
The dermatologist's opinion was taken for blisters above the right ear, which were suggestive of pyoderma, and she was given t-bact ointment to use locally. The inj. methylprednisolone was stopped after 3 days, and Tab Prednisolone was started. The patient was given soft meals and began on Sodamint and Diamox tablets. Antibiotics were not given intravenously. For pain on the surgical side of the face, gabapentin was added, and Tab. amlodipine was started i/v/o hypertension. Tab. prednisolone was gradually reduced until it was no longer needed.
He received all of the treatments and had a successful outcome. He was able to continue his normal activities after his signs and symptoms had disappeared.

Follow-up and outcomes:
The condition of the patient improved. Physiotherapy for the face is recommended. With a blood pressure in the 95th percentile and no active complaints about feedings, the patient is vitally and hemodynamically stable. As a result, the patient is discharged with a 15-day return appointment.

Discussion:
The patient was admitted to the our hospital with the complaint of hearing loss, gait instability, hazy vision, balance problems, difficulty speaking, fever, weakness, and headache. The patient was diagnosed with Neurofibromatosis type 2 after the evaluation was completed, and surgical intervention was undertaken. The patient condition was improved.“Neurofibromatosis type 2 is a genetic disorder caused by mutations in the NF2 gene on chromosome 22. It is a dominantly inherited disorder. Wishart's sickness, first diagnosed in 1822, is indeed an uncommon ailment, with an incidence of roughly 1 in 60,000." In 2013, George Sarin Zacharia[5]. Treatment is responsible for a large portion of the morbidity associated with these tumours. Most families can now be tested for the NF2 mutation, while about 20% of what appear to be random cases are actually mosaic for their gene. Inactivation of the NF2 gene product, merlin/schwannoma, contributes to the formation of both NF2 related and spontaneous tumours as a classical tumour suppressor. Gareth R Evans in the year 2001.[6]this ailment was formerly thought to be a part of von Recklinghausen's disease, but it was subsequently recognised as a unique entity with different causing genetic alterations.[7-10]
Vestibular Schwanoma is the most common phenotypic presentation, which is commonly bilateral. Vestibular Schwannoma can affect up to 95% of adult individuals with NF2.[8] However, 50% of patients with NF2 do not have a positive family history, and many have tumours or lenticular opacity before developing acoustic neuromas.Hence, The diagnostic criteria were later amended to allow for the diagnosis of NF2 in certain patient groupings. The Manchester clinical diagnostic criteria provide for the most sensitive identification of NF2 in the aforementioned subgroups.The diagnosis is confirmed by genetic tests and mutation analysis.[11-14]
The treatment of NF2 often presents a significant challenge to the treating physician in terms of surgery scheduling, type, and strategy. Despite these difficulties, surgical removal is still the preferred treatment. An expert surgical team has been demonstrated to provide a considerable mortality benefit to the patient. [15-19]

Conclusion:
Neurofibromatosis type 2 is a neurological disorder that causes noncancerous tumours to grow. The most common tumours associated with neurofibromatosis type 2 are vestibular schwannomas and acoustic neuromas. Under NIM surveillance, a 15-year-old boy with Neurofibromatosis Type 2 underwent left Retromastoid Suboccipital Craniotomy and left vestibular Schwannoma (CP angle Tumour) excision. Medical, surgical and psychological treatment works well for him.

Informed consent: Before taking this case, information was given to the child, and their mother and informed consent were obtained from the child as well as the mother.

Conflict of Interest: No conflict of Interest.
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