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Clinical Case Report

Clinical Case Report on Adolescent Onset Neurofibromatosis Type 2

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ABSTRACT

Background: Neurofibromatosis Type 2 is an inherited autosomal dominant syndrome, characterized by multiple neoplasms of the central and peripheral nervous system associated with ocular abnormalities. In this report, we aim to present the case of a 15-year old male whose chief complaints were unexplained falls, imbalance and decreased hearing. During his workup, which included cranial and whole spine magnetic resonance imaging he was found to have a Bilateral Vestibulocochlear schwannoma with multiple meningioma. Based on clinical and imaging findings, the diagnosis of neurofibromatosis type 2 was made. Our report also details the long term follow-up of a vestibulocochlear schwannoma with growth patterns before and following treatment. **Case Description:** A 15-year old, energetic male teenager, athletic and a skilled musician was diagnosed as suffering from Neurofibromatosis 2, in September 2019. At presentation, he had bilateral vestibular schwannoma, right optic nerve sheath meningioma, small anterior falx meningioma, cervico medullary and dorsal cord lesions (likely ependymomas) and multiple cauda equina lesions (likely nerve sheath tumors–schwannoma) revealed by imaging and treated by surgery over a period of 2 years. He was finally treated with stereotactic radiotherapy and at 1 year following treatment tumor showed shrinkage. He presented to our clinic for his biannual MRI of the brain with and without contrast. **Conclusion:** This case represents the sporadic occurrence of vestibular schwannomas in a young patient. The age of the patient, the presence of multiple tumors and in addition to the typical vestibular schwannomas led to a diagnosis of neurofibromatosis type 2 (NF- 2). With no known family history, the use of cranial imaging studies in addition to clinical findings, played a key role in his diagnosis. The individualized treatment provided played a significant role in tumor size shrinkage. Follow-up cranial MRI studies were significant in tracing changes in the tumor morphology.

KEYWORDS: Case Report, Neurofibromatosis Type 2, Vestibular Schwannoma, MRI

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1. Introduction

Neurofibromatosis 2 (NF2) is a slow-growing and nonmalignant, autosomal dominant inherited cancer syndrome with an estimated incidence of 1:33000 [1]. It has a penetrance of nearly 100% by the age of 60 years and there is no gender predilection. It results from a germline mutation of the NF2 tumor suppressor gene located on the long arm of chromosome 22, which encodes a tumor suppressor protein called Merlin (schwannomin). The role of merlin is to stop tumors and in its absence, cells lose the ability for contact-dependent growth arrest resulting in MISME syndrome – Multiple Inherited Schwannoma Meningiomas and Ependymomas. While the clinical manifestations of NF2 include central and peripheral nervous system tumors (nerve sheath tumors, meningioma and ependymomas), cutaneous lesions, ocular pathology and peripheral neuropathy, the hallmark of NF2 is the development of bilateral vestibulocochlear schwannoma. Patients can also have hearing loss, flesh-coloured skin flaps and muscle wasting. There can be plaque-like lesions which will be more pigmented concerning the surroundings and with increased hair, development of subcutaneous nodules located alongside the peripheral nerves [2]. Other symptoms include meningioma, ependymomas, dermatological tumors, cataracts, and renal hamartomas [2]. Bilateral vestibulocochlear schwannoma is present in 90 to 95% of NF2 patients [3].

1.1. Diagnosis

The diagnosis of Neurofibromatosis type 2 is based on physical examination, MRI results, history of hearing loss and family history. Since NF2 is a genetic and hereditary condition, there's a 50 per cent chance that the offspring of the NF2 parent will be affected. Although genetic analysis is not necessary for every neurofibromatosis case it can be considered if the diagnosis remains unclear [4]. One of the following is required to establish the diagnosis: [5]

1. Bilateral vestibular schwannoma
2. A pathogenic variant of NF2 in NF2-related tumors like schwannoma, ependymoma, meningioma etc.
3. Two major criteria
4. One major and 2 minor criteria

Major Criteria

a) Unilateral Vestibular schwannoma
b) First degree relative
c) 2 or more meningiomas
d) NF2 pathogenic variant in unaffected tissue like blood

Minor Criteria

e) Non vestibular schwannoma, ependymoma
f) Single meningioma
g) Subcapsular cortical cataract, retinal hamartoma, epiretinal membrane in patients < 40 years of age

1.2. Management

The management of NF2 involves a multidisciplinary approach which requires the involvement of oncologists, neurologists, neuroradiologists, neurosurgeons, ophthalmologists and geneticists. The mode of treatment depends on the severity of the tumor. If the schwannomas are small and asymptomatic, conservative management and follow-up with MRI is required. However, for large symptomatic schwannomas, surgery is the modality of choice [6]. Radiotherapy can be considered for non-surgical candidates for a maximum intracranial diameter of 3cm [7]. Monoclonal antibodies targeting Vascular endothelial growth factor (VEGF) like Bevacizumab have shown regression in tumor and improvement in hearing [8].

1.3. Prognosis

While improvements in care and early diagnosis are resulting in improved outcomes, many individuals with NF2 still die young.

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The presence of truncating mutations, the number of meningiomas, and the early age of development all influence the prognosis [7].

1.4. Learning Points

This case report highlights the significant challenges faced by a 15-year-old boy who was diagnosed with Neurofibromatosis type 2 without pertaining to any cue of family history. The rapid progression towards complications like schwannomas, meningiomas and spinal tumors emphasized the importance of regular and frequent monitoring through imaging and clinical assessments to prevent long-term sequelae. This case has been a challenging one as a bright budding student and guitarist undergoes sudden unexpected complications one after the other. The multidisciplinary approach by a team of specialists that included a neurologist, otolaryngologist, surgeon, radiologist, ophthalmologist and audiologist in optimizing treatment strategies and improving the overall quality of life for the patient has been integral. Moreover, the family's support in promoting a holistic environment for the adolescent has played a pivotal role in combating the psychosocial challenges and at the same time ensuring a smooth transition towards decision-making and health care. By highlighting these learning points, this report holds significance in contributing to the understanding of the complexities associated with Neurofibromatosis type 2 in adolescents and ultimately helps to improve patient care and outcomes.

2. Case Presentation

2.1. History of Patient

A 15-year-old male with principal complaints of unexplained falls, imbalance and decreased hearing. The teenager was healthy till August 2019, when he developed an imbalance while walking. The parents noticed him swaying towards the left while walking and that he frequently bumped into objects on his left side. The imbalance worsened at night and he found it increasingly difficult to go down the stairs. His symptoms were associated with occasional fine tremors while holding objects and unilateral right-sided headache. He described the pain as dull and aching, around the temporal area with neither radiation nor diurnal variation and relieved with vomiting. Within 3 weeks of the onset of initial symptoms, he developed unilateral left-sided hearing impairment. It was insidious in onset and gradually progressive and he noticed it while listening to songs. He could hear the noises but could not comprehend the words. He also held the phone to his normal right ear. His hearing did not improve in noisy surroundings. An incidental finding elicited by the physician was a history of painless, non-pulsatile proptosis of the right eye which had been present for over 2 years. As it was an innocent presentation with no additional symptoms, it went unnoticed until they presented to the ENT doctor for a hearing assessment. The proptosis remained non-progressive. There was no history of seizures or any other sensory or motor symptoms in the individual. There was no contributory past or family history.

2.2. General Examination

General Examination of the patient with normal higher psychic functions revealed an obvious right eye proptosis with an upper scleral margin. Multiple café- au-lait spots were seen on his back. A soft, nodular cutaneous swelling, (neurofibroma) of size 1x1cm was seen on the skin of the anterior abdominal wall, around his periumbilical region. His local ear examination showed normal bilateral external auditory canals and tympanic membranes but a finger rub test showed decreased hearing in both ears with the left ear affected more than the right. Pure tone audiometric testing revealed mild right-sided sensorineural hearing loss and profound sensorineural hearing loss in the left ear. His cranial nerve examination revealed that he had bilateral papilledema but he was able to count at 4 feet (VA=CF 4m) with no field defects. He had an unequal pupillary reaction with the right pupil at 4mm and the left pupil at 2 mm with no relative afferent pupillary defect (RAPD). Horizontal nystagmus was noted. His facial sensations along with the motor component of the trigeminal nerve were intact. Conjunctival and corneal reflexes were intact and he

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had no facial nerve palsy. His tongue and uvula were centralized. His motor and sensory systems tested normal for tone and power with no sensory deficits. His reflexes remained unaffected with normal cerebellar functions and no spasticity. His gait was ataxic with a positive Romberg's test (left-sided sway).

2.3. Initial MRI Evaluation

The first detailed MRI Brain (dated 10th September 2019) showed masses as documented below. It is to be noted that the volume of all masses is enumerated as Anteroposterior X Transverse X Craniocaudal. The MRI showed T2 heterogeneously hyperintense lesions bilateral cerebropontine angles with intense contrast enhancement and extension into corresponding IAC causing widening of the same (Figure 1A and 1 B). These measured 3.1X2.9X3.2cm on the right and 4.8X3.8X4.3cm (AP X TR X CC) on the left side; characteristic of bilateral acoustic schwannomas. Bilateral VII-VIII cranial nerves could not be separately visualized. Significant mass effect on the midbrain, pons and medulla from the left side and on pons and medulla from the right was noted, with a shift of brainstem to right, causing morphological distortion. Compression of the fourth ventricle with associated obstructive hydrocephalus of third and lateral ventricles – suggesting chronicity in nature due to lack of symptoms and evidence of trans-ependymal CSF seepage in T2 FLAIR images. Evans ratio measured 0.36. A right optic nerve sheath meningioma measuring 5.2X1.3X0.8cm was seen as an intensely enhancing space occupying a lesion along the lateral aspect of the right optic nerve within the orbit, extending intracranially into the lateral portion of the right cavernous sinus. The right optic nerve was of normal size with no abnormal enhancement. A mild right eye proptosis was also noted. An anterior falx meningioma was seen as a small moderate to intensely enhancing lesion in the anterior interhemispheric fissure measuring 7X5mm. The left frontal lobe, right cerebellar white matter and left posterior cerebellar margin showed T2 and T2 FLAIR hyperintense small lesions. In the left frontal lobe antero-inferiorly a cortical-subcortical T2 hyperintensity measuring 1.7X1.2X1cm was seen. Left posterior cerebellar lesion; 8X4mm was ill-defined and showed few blooming artifacts. Right cerebellar white matter T2 hyperintensity measured 8X9X4.8mm. Partial empty sella with increased CSF content within bilateral optic nerves, which showed increased vertical tortuosity and nerve head flattening and mild optic disc bulge – favouring secondary intracranial hypertension with mild papilledema.

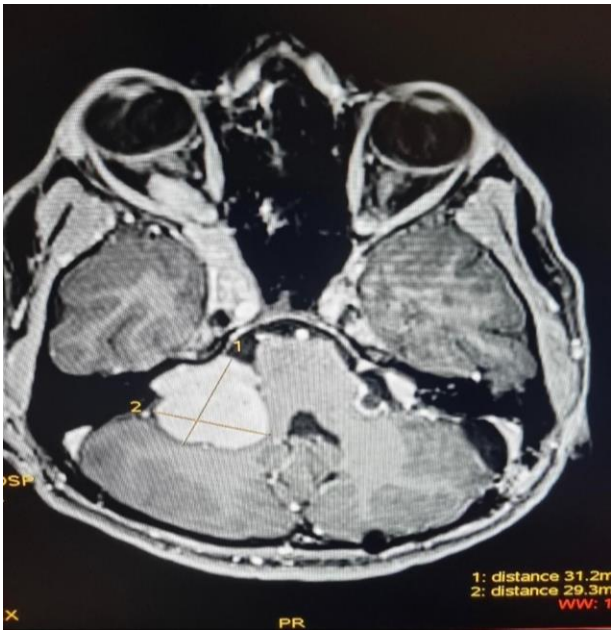


Figure 1A

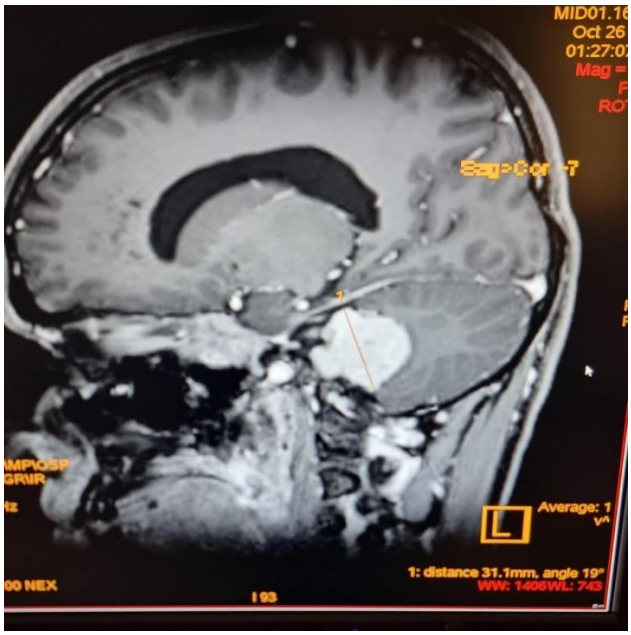


Figure 1B

Figure 1A and 1B: MRI showing T2 heterogeneously hyperintense lesions bilateral cerebropontine angles.

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The initial whole spine screening showed expansible cervico-medullary cord lesions with multiple T2 hyperintense areas and T2 hypo intense fluid levels, measuring 4.7cmX1.9cm, at C1, C2 levels. Smaller (<5mm) enhancing intramedullary lesions were also noted in the cervical cord (C4 to C6 levels) and one at D5 level. Another enhancing slightly expansible intramedullary cord lesion at the D1 level measuring 2cmX1.2cm. There were numerous sub-cent metric to tiny enhancing nodular lesions seen about the cauda equina. The bilateral acoustic schwannomas, right optic nerve sheath meningioma, small anterior falx meningioma, cervico-medullary and dorsal cord lesions (likely ependymoma) multiple cauda equina lesions (likely nerve sheath tumors – schwannomas) and cerebral and frontal lobe lesions most likely hamartoma/ dysplasia) favoured a diagnosis of NF-2 and as such with a diagnosis of NF-2, he was referred for a neurosurgeon opinion. Over a period of 4 years, the patient was offered multiple treatment modalities which included microsurgical resection, stereotactic radiotherapy and continued observation with imaging at timed intervals. The first procedure addressed the left-sided vestibular schwannoma and the cervical ependymoma. The micro-resection and decompression were done on 23rd September 2019, wherein he underwent a left retro sigmoid craniotomy and near-total tumor excision with intraoperative cranial nerve monitoring. A very small residue was left behind on the facial nerve. The facial nerve was stimulable at 0.2mA at the end of the procedure. Post-op CT brain done on the 24th Sep 2019, showed expected postsurgical changes of left sub-occipital craniotomy with right mastoid approach and resection of the left cerebellopontine angle tumor. He developed a left-sided grade 3 LMN facial nerve palsy (House and Brachmann grade 3) but his condition was stable at discharge. Post-operative specimens sent for histopathological evaluation included a CUSA trap specimen that confirmed schwannoma. As planned, he then underwent a second procedure on 4th November 2019 for plexiform schwannoma of the right ophthalmic nerve, which involved a right frontotemporal craniotomy, superolateral orbitotomy and tumor decompression. On the 5th of November 2019, in the immediate postoperative period, a plain CT Brain was done to assess the right cavernous sinus changes post-tumor excision. CT showed evidence of right frontal and left occipital craniotomy, with due changes (good decompression of the lesion) noted along the cavernous sinus. A small hyper-dense falx meningioma along the anterior interhemispheric fissure was seen. A circumscribed mildly hyper-dense lesion was also seen along the right CP angle and an expansile lesion of the cord. In the postoperative period, he remained stable with a vision at CF >4ft bilaterally with both pupils equal and reacting at 2.5mm. He had right lateral rectus paresis which gradually improved but his facial paresis persisted. He was discharged with due medications which included Levetiracetam. Soon after, on 5th December 2019, he developed Levetiracetam-induced psychosis requiring hospitalization for aggressive and anxious behavior. He was treated with antipsychotics and upon improvement of symptoms was discharged with advice to continue medications. He once again required hospitalization for psychosis on 1st January 2020 and the same management was given. His initial audiometric evaluation on 20th January 2020 showed right-sided mild sensorineural hearing loss and left-sided profound hearing loss. His speech discrimination scores were very poor on the left side and 65% on the right and the same persisted in subsequent audiometric evaluations. Audiometry findings taken on 16/06/2020 showed right-sided mild sensorineural hearing loss and left-sided profound hearing loss (Figure 2).

The patient decided not to undergo further surgical treatments and was followed up with repeat MRI imaging done at regular intervals to note changes in tumor morphology as documented below:

2.4. Follow-up MRI of the Brain & Spine on 07/01/2021

(In comparison with previous MRI dated 10.09.2019) – The right acoustic schwannoma was slightly enlarged compared to previous MRI, measuring – 3.9cmX3.8cmX3.2cm. No diffusion restriction was seen. Significant mass effect on right side of pons and medulla with a shift of brainstem and fourth ventricle to left. Compression of the fourth ventricle with partial effacement was noted. No significant hydrocephalus was seen and Evans ratio measured to be 0.29. The residual lesion in the left CP angle and IAC acoustic schwannoma, measured 1.1cmX0.6cmX1.1cm. An intensely enhancing space-occupying lesion along the

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lateral aspect of the right optic nerve 'within the orbit, extending intracranial into the lateral portion of the right cavernous sinus – favouring optic nerve sheath meningioma remained similar to the previous MRI. The right optic nerve was of normal size and showed no abnormal enhancement. The right eye proptosis was reduced. Small falx meningioma of the anterior interhemispheric fissure remained similar. The left frontal lobe, right cerebellar white matter and left posterior cerebellar margin showed T2 and T2 FLAIR hyper-intense non-enhancing small lesions which remained similar. Partial empty sella is noted with increased CSF content within bilateral optic nerves. The major intracranial arteries and intracranial venous sinuses did not show obvious abnormality. Expansile cervico medullary cord lesion with multiple T2 hyperintense areas, favouring ependymoma, showed a slight increase in size, now measuring 5.5cmX1.9cm, at C1, C2 levels. Smaller multiple <5mm enhancing intramedullary lesions were seen in the cervical cord (C4 to C7 levels) and one at D5 level. Another enhancing slightly expansile intramedullary cord lesion at the D1 level remained similar in size. Numerous sub-centimetric to tiny enhancing nodular lesions are noted about the cauda equina.

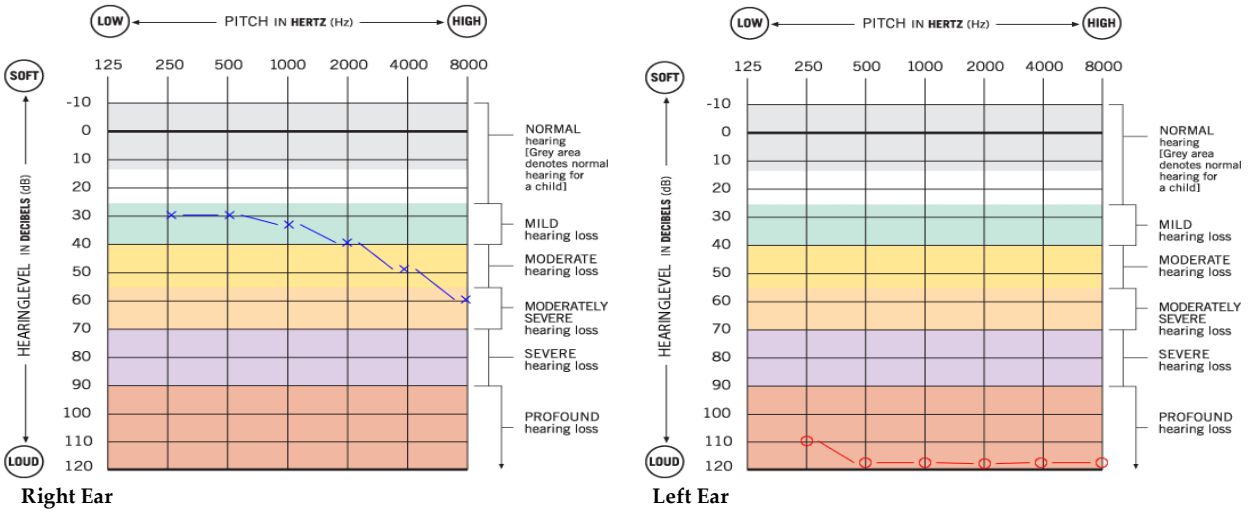


Figure 2. Audiometry findings dated 16 June 2020.

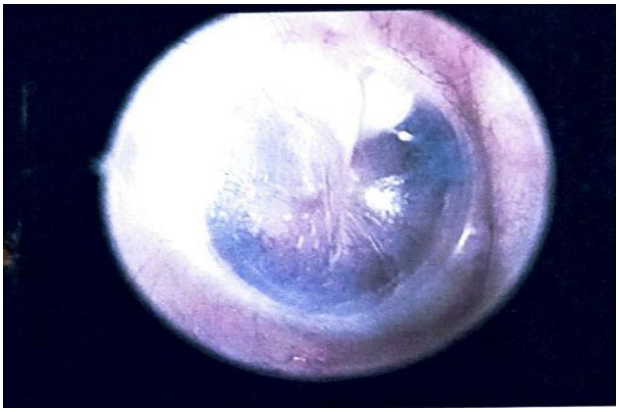
22/05/2022 shows: The right-sided CP angle mass measured 31X35X32 mm (was 46X45X41mm) and the left-sided CP angle mass 13.5X15X21 mm (was 9X15X15mm). The right intra-orbital meningioma measured 15X19X21mm (was 12.6X19mm). The upper intra-medullary upper cervical cord mass (at the cervico-medullary junction) was 20x20X77 mm (was 19X64 mm). The lower cervical cord intra-medullary mass measured 13.5X31 mm (was 15X30 mm). The foci of cord swelling at the level of D5 and D10 vertebrae levels (intra-medullary small masses) showed mild progression in size with central enhancement; at the level of the upper aspect of the D5 vertebra the mass was measured 5X10 mm and at the level of D10 vertebra was 7X14 mm). The dumbbell-shaped schwannoma at the right C5/ 6 vertebral level remained unchanged. Interval minimal enlargement of the multiple enhancing mass lesions along the thoracic cord and the cauda equina, the largest spherical-shaped mass at the level of the D12 vertebra measured 16 mm. Otoendoscopy conducted on 06/07/2022 showed the right tympanic membrane to be intact while the left tympanic membrane was thin intact (Figure 3).

He was started on Stereotactic Radiotherapy for the Right-sided Vestibular Schwannoma on 15/06/2022 and the course continued for a month up to 20/07/2022. He was planned for 5040 cGy in 28 fractions, however, due to logistic issues one fraction was skipped and he received a dose of 48.6Gy in 27 fractions (180cGy per fraction, 5 Fractions per week] using 4 arcs of 6 MV photons. The dose prescribed to 100% isodose line. He tolerated the treatment very well without any major side effects or interruptions. No neurologic deficits or count drop occurred and there was no need for steroids throughout the course. Some

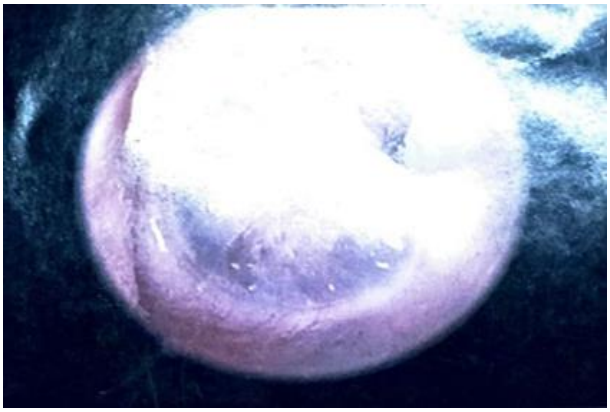
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of the masses showed mild progression, some remained stationary and some showed mild regression. No definite interval of new masses. Follow-up MRI after radiotherapy on 28/12/2022 -The mass in the right CP angle measured 33X35X30 mm, (was 31x35x32 mm). The mass in the left CP angle measured 15X18X18 mm, (15x19X21mm). The right orbital mass measured 17X19X25 mm, (15X19X21 mm). The right-sided intra-osseous meningioma measured 14X22X29 mm (was much smaller in the previous scan) with the possibility of engulfing the right internal carotid artery and right cavernous intra-dural venous sinus. The anterior falx small meningioma remained the same. The intra-medullary upper cervical and medullary mass measured 20x23x77mm, (20X20 X77mm). The lower cervical cord intra-medullary mass measured 15X37 mm, (was 13.5 X 31mm). The right-sided dumbbell schwannoma at the level of C5/6 vertebrae IV foramen measured 14X24 mm, (12X12 mm). The left-sided dumbbell schwannoma at a level in the intervertebral foramen of D1 vertebrae was 24X28mm, (was 20 X 16 mm). The spherical solid mass at the level of the D12 vertebra (measuring 17mm) remained unchanged. The extensive small intra-medullary tiny masses and the mass on the roots of the cauda equina remained the same. The latest MRI study dated 30/5/2023 in comparison to the previous scan dated 28.12.2022. The mass in the right CP angle is currently measuring 33.6X32.6X27 mm, (was 33X35X30 mm) and the mass in the left CP angle currently measures 20X18X14 mm (was 15X18X18 mms). The right orbital mass is now 13X35X16.6 mm (was 17X19X25mm), showing interval enlargement. The right-sided intra-osseous meningioma is still 14X22X29 mm and is still engulfing the right internal carotid artery and right cavernous intra-dural venous sinus. The anterior falcine small meningioma is the same at 7 mm. The intra medullary upper cervical and medullary mass is currently measuring 20X23X77 mms in dimensions (not significantly changed). The lower cervical intra medullary mass is currently measuring 15X37 mm and unchanged. The right-sided dumbbell schwannoma at the level of C5/6 vertebrae IV foramen is still currently measuring 14X24 mm dimensions unchanged. The left-sided dumbbell schwannoma at the level of the intervertebral foramen of the D1 vertebra is currently measuring 32X36 mm in dimensions (interval enlargement, was 24X28 mm in dimensions. Thus after a year some of the masses are showing mild progression, some are stationary and some are showing mild regression. No definite interval of new masses. The patient, to date, is less inclined to surgical interventions as he has no new masses and even the masses with mild progression do not elicit new clinical symptoms.



RT Tympanic Membrane Intact



LT Tympanic Membrane Thin Intact

Figure 3. Tympanic membrane findings.

2.5. Restoration of hearing loss

As a budding guitarist, he was hard hit by the loss of his hearing which exceeded a level that could be rehabilitated by hearing aids, cochlear and brainstem implants. He strongly desires the return of his auditory faculty and his recent consultation was specific in the pursuit of the bone-conduction hearing implant (BCHI). But he was disillusioned at the advice offered. He was advised to undergo surgery and micro-resection of the CP tumors before considering auditory implants. All BCHIs contain permanent magnets, and MRI compatibility with a BCHI is a challenging feat. This is more so in his case, as he opted

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out of surgical interventions. Some of his tumors show progression, albeit mild; and one must also not forget the risk of malignant transformation. As such he needs continued observation with imaging and is thus heavily dependent on MRI scans at timed intervals. If such imaging is obtained with the magnetic device implanted, many adverse effects could occur. MRI performed on patients with an implanted magnetic device could potentially result in a migration of the device. Demagnetization and malfunction of the devices could also occur, and the heating of such internal devices could damage the surrounding tissues. These effects can be avoided by removing the magnets before performing MRI scans or by using Low-Tesla (T) MRI [9].

2.6. Quality of Life

The central goal in the patient's management was to address the tumor load and maintain normal functions in addition to ensuring a quality of life. He is managed by a multidisciplinary team involving a physician, neurosurgeon, otolaryngologist, ophthalmologist, neurologist, radiologist, pathologist and audiologist. With an early diagnosis and prompt surgical interventions, his tumor load was considerably arrested and he remains asymptomatic. He experiences social and emotional difficulties, including anxiety, depression, low self-esteem and/or body image, social withdrawal, and behavioral problems. His diagnosis had come at a crucial period of his high school and there was a considerable hiatus in his academics, which he overcame by sheer will and despite the difficulties in school he cleared his boards and joined college in August. He was pulled out of college within a month as he was struggling to cope and finding it extremely difficult to adjust both physically and emotionally. He now continues his pursuit of knowledge through online courses. With our case study, we have concluded the challenges involved in the care of a young NF-2 patient. This patient has done well overall secondary to the care provided to him by all of his providers. He will continue routine surveillance whether or not he is symptomatic from tumors.

Declaration:

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Informed Consent

Informed Consent was obtained from the patient before the writing of the manuscript.

Author Contributions

Authors Afrah and Kevin collected the data from author Dr. Nadeeja and wrote the clinical case of the paper. The literature review was handled by authors Dr. Fatema and Aysha. The final paper was proofread by all the authors mentioned here.

Data Availability

The authors declare that data supporting the findings of this study are available within the article.

Conflict of interest

The authors declare no conflict of interest.

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