

Case study

CASE REPORT ON RIGHT ACOUSTIC AND LEFT TRIGEMINAL SCHWANNOMA , HYDROCEPHALUS WITH NEUROFIBROMA A RARE CASE

ABSTRACT

A vestibular schwannoma or acoustic neurinoma, or acoustic neurilemoma is a slow growing benign tumor arises balance and hearing nerves in the inner ear. It is caused by overabundance of Schwann type of cell, which support and insulate nerve fibers, wrap onion skin around them. The nerves that control hearing and balance are impaired when vestibular schwannoma increases in size, resulting in hearing loss that is one-sided or asymmetric, tinnitus and loss of balance. When a tumor develops large enough, obstruct the facial nerve, resulting in numbness in the face. Vestibular schwannomas can also damage the facial nerve, resulting in facial weakness or paralysis on the tumor's side. If the tumor becomes large enough, it press against surrounding brain areas like the cerebellar and brainstem, posing a life-deteriorating hazard.(1) 17 years old male child was admitted in neuro ward with rare case of right acoustic and left trigeminal schwannoma, hydrocephalus with neurofibroma. In the present case, the treatment approach was mainly underwent in the form of right V.P.Shunt done and treated with antibiotics, antacids, anticonvulsants, analgesics, brain stimulants, protectants, multivitamins and other supportive treatment. Nurses have to play an important role to identify such type of symptoms and they should think critically, take action immediately to provide care to such type of patients.

Keywords: Vestibular neuroma, acoustic and trigeminal schwannoma, neurofibroma, meningioma, hydrocephalus

CASE REPORT:

A 17 years old male was admitted with the diagnosis of right acoustic and left trigeminal schwannoma, hydrocephalus with neurofibroma with present complaints of headache, vomiting and visual disturbance since two month and also history of taken creatinine previously during gym training. On central nervous system examination conscious, obeyed vocal command, weakness present in lower limb. He was investigated radiologically and hematologically. MRI done- right basi-occipital extra axial enhancing lesion measuring 1.8 x1.6 cm appears meningioma right CP angle enhancing nodular lesion in left measuring 9 x 8mm appears small acoustic schwannoma another similar lesion in left retroseller plane measuring 1.5 x1.4cm abutting optic chiasma anteriorly and enhancing bifurcation of left ICA appears small meningioma schwannoma and mild ventriculomegally probably early hydrocephalus. MRI cervical spine C3 to C6 right sided intraspinal extramedullary oblong lesion measuring 4.2 cm in length.1.0 min AP

compressing the cervical cord which is displaced towards right and moderately compressing spinal and markedly compromising right lateral recess appears meningioma probably components of neurofibromatosis. In the form of MRI brain findings, MRI LS spinal reveals L1-S2 intraspinal lobulated enhancing mass lesion subtotally obliterating the spinal canal may represent maxillary ependymomas component of neurofibromatosis in view of MRI brain and cervical spine finding. His haemogram, renal profile, liver enzymes, BSL, serology test, coagulation profile were normal. Patient underwent surgery in the form of right ventricular peritoneal shunt and treated with antibiotics, antacids, anticonvulsants, analgesics, brain stimulants, protectants, multivitamins and other supportive treatment

Discussion:

As seen in the top image, acoustic neuroma is a tumor that forms in a benign manner on the balancing and cochlear nerves that connect inner ear to your brain. Hearing loss and unbalance may result from the tumor's pressure on the nerve.¹ Schwannomas are benign tumors that develop in the nerve sheaths of cranial nerves that run alongside the cerebellum and brainstem. Vestibular Schwannomas are the most prevalent, causing tinnitus and loss of hearing. It grows larger, can affect incoordination, imbalance also develop facial paralysis.⁽²⁾ Acoustic neuromas have a mainly unknown source. There has been no scientific proof that any environmental factor (such as cell phones or nutrition) causes malignant cancers. Acoustic neuromas can be random or be caused by neurofibromatosis type 2 (NF2), an inherited disorder (NF-2). 95 percent of tumors are sporadic, while only 5% of auditory neuromas are caused by NF-2. Neurofibromatosis is an uncommon disease that can manifest itself in two ways. Tumors occur on nerves all over the body, especially in the skin, as a result of Type 1. Type 2 acoustic neuromas can develop on both the left and right sides, posing the risk of total hearing if the tumors progress unchecked. Because hearing preservation is a primary goal, the occurrence of bilateral acoustic tumors influences treatment options.⁽³⁾ Most symptomatic schwannomas can now be treated with minimally invasive keyhole and endoscopic surgery. The most frequent are vestibular schwannomas, which cause ringing in the ear and loss of hearing; as they grow larger, can affect incoordination, facial paralysis and imbalance. Numbness, tingling, or pain in the face is symptoms of trigeminal schwannomas, which are less common. Maximal safe surgical removal is the majority of symptomatic vestibular and trigeminal schwannomas respond well to this treatment. A retro mastoid keyhole craniotomy can be used to remove most large acoustic neuromas, whereas a retro mastoid approach can be used to remove most trigeminal schwannomas.²

Vestibular schwannomas (VS) form in the internal auditory canal, cerebellopontine angle, cochlear, and labyrinth, and grow slowly and push the out of brainstem and crush it. Vestibular schwannoma accounts for 6–7% of intracranial tumors, incidence of annual 1:100,000. They are by far the most neoplasm in the cerebellopontine angle, accounting for 90 percent of all lesions. The first section of this research covers tumor stages, the most common grading schemes for facial nerve function and hearing, and a quick description of vestibular function tests.⁽⁴⁾

Bilateral auditory neuromas may be a symptom of neurofibromatosis II type, which is due to a II gene neurofibromin mutation, which codes for protein merlin, on chromosome 22q12.2. Acoustic neuroma has a causal propensity mutation, according to studies. A patient's exposure to

radiation may also predispose them to developing that ailment.(5) Despite the fact that radiation of mobile phone a source of concern, multiple failed researches to show that it is the cause of vestibular schwannomas..(6) Schwannomas are responsible for about 8% of all cerebral tumors that present clinically. Mostly scattered and unilateral are acoustic neuromas. Neuromas both sided auditory hereditary nature account for schwannomas less than 5%. Acoustic schwannomas can occur in children, albeit they are uncommon. There is a slight female preponderance, with issues worsening during pregnancy. Hereditary auditory neuroma is substantially more common in NF II than in NF I, despite the being far common. In 24 percent of individuals with NF I, unilateral auditory neuroma has been recorded exclusively. In NF I, the faulty genetic locus was found on chromosome 17; in NF II, it was found on chromosome 22.(7)

Trigeminal schwannomas make up 0.07 percent to 0.36 percent of all intracranial tumors, and 0.8 percent to 8% of all intracranial schwannomas. Understanding the natural history of tumor's and outcomes of treatment is necessary for selecting the best management strategy. A 36 year old man chief complaint with severe headache, imbalance, insomnia and dizziness during a three-month period is described in this article. Magnetic resonance imaging revealed a significant enhancing lesion centred on the left Meckel's cave and spreading into both the middle and posterior fossas in patients with obstructive hydrocephalus caused by compression of the fourth ventricle. The tumor's posterior fossa component was resected to ease the effect of mass on the without brainstem trying a drastic resection of the middle fossa component, which could result in severe cognitive impairment. Trigeminal schwannoma was confirmed diagnosed by a histological examination. After a subtotal surgical excision, the remaining tumor demonstrated progressive volumetric decrease. This example demonstrates the importance of a scheduled operation in schwannomas, as well as the difficulties in evaluating effect of treatment surgically or with stereotactic radiotherapy on benign tumor.(8)

Conclusion:

The right acoustic and left trigeminal schwannoma, hydrocephalus with neurofibroma is a rare disease. Immediate surgical intervention ventriculoperitoneal shunt done and treated with antibiotics, antacids, anticonvulsants, analgesics, brain stimulants, protectants, multivitamins and other supportive treatment, will go a long way in helping such children to lead a normal life.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard guideline patients consent and ethical approval has been collected and preserved by the authors.

Reference:

1. Vestibular Schwannoma (Acoustic Neuroma) and Neurofibromatosis [Internet]. NIDCD. [cited 2021 Jul 13]. Available from: <https://www.nidcd.nih.gov/health/vestibular-schwannoma-acoustic-neuroma-and-neurofibromatosis>

2. Schwannoma (Trigeminal & Acoustic) [Internet]. Pacific Brain Tumor Center. [cited 2021 Jul 13]. Available from: <https://www.pacificneuroscienceinstitute.org/brain-tumor/conditions/schwannoma-trigeminal-acoustic/>
3. Acoustic Neuroma, vestibular schwannoma | Mayfield Brain & Spine [Internet]. [cited 2021 Jul 13]. Available from: <https://www.mayfieldclinic.com/pe-acoustic.htm>
4. Rosahl S, Bohr C, Lell M, Hamm K, Iro H. Diagnostics and therapy of vestibular schwannomas – an interdisciplinary challenge. *GMS Curr Top Otorhinolaryngol Head Neck Surg*. 2017 Dec 18;16:Doc03.
5. Zhao F, Yang Z, Chen Y, Zhou Q, Zhang J, Liu J, et al. Deregulation of the Hippo Pathway Promotes Tumor Cell Proliferation Through YAP Activity in Human Sporadic Vestibular Schwannoma. *World Neurosurg*. 2018 Sep;117:e269–79.
6. Pettersson D, Mathiesen T, Prochazka M, Bergenheim T, Florentzson R, Harder H, et al. Long-term mobile phone use and acoustic neuroma risk. *Epidemiol Camb Mass*. 2014 Mar;25(2):233–41.
7. Greene J, Al-Dhahir MA. Acoustic Neuroma. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 [cited 2021 Jul 13]. Available from: <http://www.ncbi.nlm.nih.gov/books/NBK470177/>
8. Jusue-Torres I, Martinez-Gutierrez JC, Elder B, Olivi A. Giant Trigeminal Schwannoma Presenting with Obstructive Hydrocephalus. *Cureus*. 2015 Nov 20;
9. Acharya, Sourya, Samarth Shukla, Amol Andhale, Akhilesh Annadatha, and Yash Gupte. “Neuroleptic Malignant Syndrome (NMS) after Treatment with Metoclopramide - A Rare Case Report.” *JOURNAL OF EVOLUTION OF MEDICAL AND DENTAL SCIENCES-JEMDS* 9, no. 41 (October 12, 2020): 3065–66. <https://doi.org/10.14260/jemds/2020/671>.
10. Annadatha, Akhilesh, Sourya Acharya, Samarth Shukla, Amol Bhawane, Anuj Varma, and Amol Andhale. “Multiple Myeloma Presenting as Hypercalcemic Crisis: A Case Report.” *MEDICAL SCIENCE* 24, no. 105 (October 2020): 3148–52.
11. Lahole, Swapnil, Sourya Acharya, Sanket Bakshi, Samarth Shukla, and Vidyashree Hulkoti. “Acute Disseminated Encephalomyelitis (ADEM) versus Multiple Sclerosis (MS)- A Diagnostic Challenge in an Adult.” *JOURNAL OF EVOLUTION OF MEDICAL AND DENTAL SCIENCES-JEMDS* 9, no. 21 (May 25, 2020): 1672–74. <https://doi.org/10.14260/jemds/2020/367>.
12. Panchbhai, Aarati, and Rahul Bhowate. “MRI Evaluation of Involvement of Parotid and Submandibular Glands by Tongue Squamous Cell Carcinoma.” *ORAL ONCOLOGY* 102 (March 2020). <https://doi.org/10.1016/j.oraloncology.2019.104557>.

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