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A Case Report On The Management Of A Patient With Neurofibromatosis-II With Pseudo Meningocele

Rohini Moon[1], Switi Jawade[2],Aniket Pathade[3], Pragati Alnewar[4]
1] GNM 3rd year, Florence Nightingale Training College Of Nursing, Sawangi (M),Wardha, Email: rohinimoon23@gmail.com ,7774917239
2] Nursing Tutor, Florence Nightingale Training College of Nursing, Sawangi (M), Wardha India, Email: vanshikalohave19@gmail.com , 8378975256
3] Research Consultant, Department of Research and Development, Jawaharlal Nehru Medical College, Datta Meghe Institute of Medical Sciences, Wardha, Email: aniketpathade@gmail.com
4] Department of Medical-Surgical Nursing, Smt. RadhikabaiMeghe Memorial College of Nursing, Datta Meghe Institute of Medical Sciences, Sawangi, Wardha, Maharashtra.

Abstract:
Introduction: Neurofibromatosis -II is a genetically inherited condition characterized by the development of several central nervous system tumors, the most common of which are vestibular schwannomas with or without peripheral symptoms such as cataracts or cutaneous neurofibromas. NF2, unlike its type 1 sibling, is a rare condition. We present a classic neurofibromatosis type 2 with a wide range of clinical signs and typical neuroimaging findings.
Information about the patient: In tertiary rural hospital, a male child was admitted with a chief complaint of severe headache since one month, difficulty in sitting, the problem with balancing, ringing in the ear, backache etc.
Main symptoms and important clinical findings: In tertiary rural hospital, a male child was admitted with a chief complaint of severe headache since one month, difficulty in sitting, problem with balancing, ringing sensation in the ear, backache etc. Patients underwent various investigations such as complete history taking, physical examination, complete blood count, CT scan, MRI, X-Ray, and lumbar puncture all investigations are done.
Conclusion: Neurofibromatosis type 2 with pseudo meningocele is caused because of the mutation in the gene for a protein that regulate a growth of nerve tissue. After getting treatment as per doctors prescribed patient symptoms has minimize.

Keywords: Vestibular Schwannoma, neurofibromatosis type 2, meningioma, pseudo meningocele

Introduction:
The development of bilateral vestibular schwannomas is a hallmark of neurofibromatosis type 2 (NF2). Mutations in the NF2 gene on chromosome 22 cause this dominantly inherited disease. [1] We present a prototypical instance of NF2 and provide a brief review of the literature on this uncommon illness. Multiple neoplasms of the nerve system, both central and peripheral are connected with ocular abnormalities in neurofibromatosis type 2.[2] The vestibulocochlear Schwannoma is the most prevalent tumour linked with the condition, Neurofibromatosis type 2 affects 10% of people with this tumour. A 15 years old male child having a chief complaint of severe headache, difficulty in sitting, problem with balancing, ringing sensation in the ear, headache etc. A child has undergone various investigations such as MRI, lumbar puncture, X-Ray, CT scan etc. Neurofibromatosis type 2 with pseudo meningocele. The neurological disorder neurofibromatosis type 2 is characterised by the formation of non-cancerous tumours. The most frequent auditory neuroma linked with neurofibromatosis type 2 vestibulocochlear Schwannoma . the nerves that transmit the indications and symptoms expand. Neurofibromatosis Type 2 neurofibromatosis usually appears in the early twenties of puberty.[3], however, occur at any age. Hearing loss, ringing in the ears, and balance problems are the most prevalent early signs and symptoms of vestibular Schwannoma. By 30, this tumour is most commonly found in both ears. The signs and symptoms of a tumour forming anywhere in the nervous system will vary depending on where the tumour is located. Fluid accumulation in the brain, vision problems, numbness or weakness in the limbs and legs are all possible complications of tumour growth. Some persons with neurofibromatosis type 2 experience lens clouding in one or both eyes. The genetic disorder neurofibromatosis type 2 (NF2) is usually associated with bilateral vestibular schwannomas, also known as acoustic neuromas. These benign (noncancerous) tumours develop on balance and hearing nerves that lead to the inner ear. Pseudomeningoceles are aberrant collections of cerebrospinal fluid (CSF) that develop due to trauma or surgery that causes leakage from the CSF-filled areas around the brain and spinal cord. A pseudo meningocele is an abnormal accumulation of cerebrospinal fluid that develops as a result of cerebrospinal fluid leakage from areas surrounding

the brain or spinal cord as a result of trauma or surgery. In less than 2% of patients following laminectomy or discectomy, spinal pseudo meningocele can develop. A pseudomeningocele is an abnormal collection of cerebrospinal fluid (CSF) around the brain or spinal cord that interacts with the CSF space. In contrast to a meningocele, in which the fluid is encircled and limited by the dura mater, the fluid in a pseudomeningocele is contained in a cavity inside the soft tissues.

Patient information: In tertiary rural hospital a male young child 15 years admitted with a chief complaint of difficulty in sitting, problem with balancing, severe headache since one month, ringing sensation in-ear, weakness , backache etc.

Primary concern and symptoms of the patient: A male child was admitted to tertiary rural hospital in the pediatric ward with a chief complaint of severe headache since 1 month, backache, balance problem, ringing sensation in the ear , difficulty in sitting etc. At the time of admissions, the patient's principal symptoms were observed.

Medical, familial, and psychosocial background: A male child has a history of type 2 neurofibromatosis with pseudo meningocele.A male child takes a treatment for that condition and a male child belongs to a Nuclear family, and he maintain a good relationship with health professionals such as nurses, doctors, and other patients.

Relevant past intervention with Outcome : A male child was admitted to tertiary rural hospital in the paediatric ward with a chief complaint of severe headache since one month, the problem with balancing, backache, ringing sensation in the ear, and difficulty in sitting. After all investigations such as MRI, Lumbar puncture, and X-Ray doctor diagnosed a case of Neurofibromatosis type 2 with pseudo meningocele was noticed, and the outcome of the condition was good.

Clinical Findings: A male child was in good condition. In a physical examination of the patient, the spine is abnormal, difficulty in balancing the body, and other investigation such as CT of the brain. This plane identifies that VP shunt noted passing to the burr in the left parietal lobe with its tips of the septum pellucidum. In complete blood count, Haemoglobin is 12.6 gram, MCHC 33.9, MCV 22.9, platelet count is 33.9, red blood cell count is 4.05, white blood cell count is 6400, and other investigation. Such as coagulation, profile test LFT, KFT, potassium, magnesium and urine analysis. The patient vital signs are typical. This male child has no history of medical problems. Patient's condition was good. A 15 yr child was discharged after the procedure. MRI, CT scan, and spine examination during the routine paediatric follow-up found that there is some abnormality in the spine and nervous system.

Diagnostic Assessment: Patient diagnosed with a case of Neurofibromatosis type 2 with pseudo meningocele is interpreted depending on Physical examination, and other investigations are normal based on the patient's history. And surgical management of the deformity of a patient is a cure. The patient does not experience various challenges during diagnostic evaluation

Diagnosis:

A case of Neurofibromatosis type 2 with pseudo meningocele was discovered after a physical examination and study.

Therapeutic Interventions:

A patient undergone presently medical management such as Antibiotic, antipyretic, Nutritional supplements, antiepileptic medication, amino glycoside antibiotic, vasodilators given. Patient surgery of Left VP shunt Placement is done and given supportive therapy to the patient.

Follow Up and Outcome: The patient condition was improved.

Nursing Perspective:

Monitor vital signs per hour, monitor the child's heart rate, maintain intravenous fluid of patient, and maintain intake and output of the patient.

Discussion:

A male child was admitted to tertiary rural hospital in the pediatric ward with a chief complaint of severe headache since one month, the problem with balancing, backache, ringing sensation in the ear, and difficulty in sitting. After all investigations such as MRI, Lumbar puncture, and X-Ray doctor diagnosed a case of Neurofibromatosis type 2 with pseudo meningocele was noticed, patient had given medical management such as Antibiotic, antipyretic, Nutritional supplement, antiepileptic medication, amino glycoside antibiotic, vasodilators and the outcome of the condition was good.Unilateral hearing loss is the most common clinical sign in adults with NF2. Accompanying Seizure, headache, muscle weakness, paresthesia, and cutaneous tumour are some of the less typical symptoms at first presentation, as are tinnitus and tinnitus preceding hearing loss. On the other hand, cutaneous seizure

abnormalities and skin characteristics are rarely addressed. NF2 sufferers, on the other hand, are prone to cutaneous lesions.[5]

According to Asthagiri and Collegeous' 2009 study of NF2 clinical manifestations, cutaneous tumours occur in 59-60% of patients. Skin plaques, subcutaneous tumours, and intradermal tumours are all examples of NF2. They also point out that Up to 48 per cent of individuals have subcutaneous tumours and skin plaques, with intradermal tumours being less prevalent. MRI scans of the brain revealed multiple giant tumours located supra- and infratentorial at the time of admission to the hospital. According to histology, the operated tumours had bilateral acoustic Schwanoma in the cerebellopontine angles and mixed meningioma in the others.[6]

According to clinical criteria for neurofibromatoses, the patient was diagnosed with neurofibromatosis type 2 (NF2). developed weakness in all of her limbs a few months after the last operation. An intramedullary tumour in the cervical area and other localized lesions throughout the central canal were discovered on spinal MRI. Because of the clinical improvement after dexamethasone treatment and the lesion placement in the cervical medulla, surgical therapy was not used. Our example demonstrates that in NF2, there is a common lack of neurocutaneous alterations and a late onset of substantial neurological symptoms.[7]

The treatment of NF2 often presents a significant challenge to the treating physician regarding surgery scheduling, type, and strategy. Despite these difficulties, surgical removal is still the preferred treatment. It has been discovered that surgical care by an expert team confers a considerable mortality benefit to the patient.[8] However, even in the hands of a skilled surgeon, significant complications such as complete hearing loss and facial nerve injury can occur. Radiation therapy or experimental therapeutic modalities may be tried for patients who are poor surgical candidates or who refuse surgery. Because NF 2 is an autosomal dominant condition, children of affected parents should be frequently checked for phenotypic symptoms of the mutation. Examination of the nervous system, and ophthalmologic evaluation, however, 50% of patients with NF2 do not have a positive family history, and many have tumours or lenticular opacity before developing acoustic narrow.[9-14]

As a result, diagnostic criteria were changed to allow for the diagnosis of NF2 in specific patient groupings. The Manchester clinical diagnostic criteria allow for the most sensitive identification of NF2 in the subgroups mentioned above. Hearing loss and tinnitus are the most common clinical manifestations. Other signs and symptoms include dizziness, headaches, blurred vision, and facial numbness. The craniospinal tumours can be seen via neuroimaging [computed tomography or magnetic resonance imaging (MRI)].[10] Genetic tests and mutation analysis confirm the diagnosis. [15-17]

Conclusion:

Patients with neurofibromatosis frequently have a low mineral density due to dystrophic alterations, and their bones are brittle...In this situation, lengthy fusion during the initial operation may have aided in forming a solid fusion of frail bone.

Skull-based pseudo meningocele develops due to a neurologic treatment and can become problematic as they grow. Although most of these instances can be treated without surgery, those who do not respond to conservative treatment should be evaluated for surgical intervention. Patients with NF2 may have a single cutaneous Schwanoma with no other symptoms or clinical signs. In young individuals who arrive with a peripheral Schwanoma despite the absence of other clinical symptoms, further evaluation is required.

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