

Case Report

Imaging Findings for Bilateral Giant Vestibulocochlear Schwannoma

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Abstract

Neurofibromatosis type 2 mostly develops with multiple neoplasms of the central and peripheral nervous system and is associated with ocular abnormalities.

The presented case is a 19-year-old female patient with bilateral vestibulocochlear schwannomas in both pontocerebellar corners, intradural intra-extramedullary masses, and multiple neurofibromas in the spinal canal.

The clinical picture for NF-2, also called central neurofibromatosis, is completely different from von Recklinghausen disease. Untreated bilateral vestibulocochlear schwannoma may cause hydrocephalus in NF-2, and lead to death. Therefore, it is recommended to carefully monitor and treat bilateral vestibulocochlear schwannoma in accordance with its stage.

Keywords: Bilateral giant vestibulocochlear schwannoma, hydrocephalus, neurofibromatosis type 2

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Introduction

Neurofibromatosis type 2 mostly develops with multiple neoplasms of the central and peripheral nervous system and is associated with ocular abnormalities.¹ This disease affects males and females equally, and its incidence is 1/37,000.²

Bilateral vestibulocochlear schwannomas are encountered in patients with NF-2. The clinical picture for NF-2, also called central neurofibromatosis, is completely different from von Recklinghausen disease. While vestibular schwannoma is rarely seen in NF-1, the presence of bilateral vestibulocochlear schwannoma is one of the diagnostic criteria for NF-2. Diagnosis of neurofibromatosis type 2 may be difficult in the absence of cutaneous manifestations.³

NF-2 is very rare. According to our literature review, this is the first case where bilateral giant vestibulocochlear schwannomas have caused hydrocephalus that led to death. Therefore, in this report, we decided to present the magnetic resonance imaging (MRI) and computerized tomography (CT) findings.

Case Report

A 19-year-old female patient presented with symptoms of severe headache over the past two months, hearing problems in the past year, and sporadic syncope. Masses in both pontocerebellar corners were identified using an MRI scan. Mild dilatation of the lateral ventricles secondary to mass pressure was also observed. Furthermore, intradural intra-extramedullary masses and multiple neurofibromas extending into the parasagittal tissues from the neural foramina were noted in the spinal MRI scan.

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Additionally, three neurofibromas were identified in hairy skin, but cafe-au-lait stains were not encountered. Ocular findings were normal. In the audiological study, total hearing loss on the left and advanced sensorineural hearing loss on the right were identified. The patient was diagnosed with NF-2 based on the clinical and imaging findings. Her family history was not remarkable. Due to the intracranial mass, surgery was recommended for the patient. However, the patient did not agree to the treatment; thus she could not be operated on. Two months later, the patient presented to our hospital again due to the sudden loss of consciousness. According to the neurological examination, her overall condition was poor; she was confused in terms of her consciousness, and had a rating of 9 on the Glasgow coma scale. Emergency brain CT and brain MRI scans were performed. According to the cranial CT scan, massive lesions, which were evidently widening the acoustic canals in the bony window, were exerting significant pressure on the brain stem and cerebellum on both pontocerebellar corners causing hydrocephalus (Figure 1). Subsequently, contrast cranial and spinal MRI scans were taken using the Siemens Symphony 1.5 T MRI device. During the MRI scan of the brain, axial T1A, T2A, FLAIR, sagittal 2A, as well as contrasted axial and coronal sequences were received. During the MRI scan of the spine, axial and sagittal T2A, T1A, as well as contrasted axial and sagittal T1 AG sequences were received. In the cranial MRI, hypo-intensity in T1-concentrated sequences extending from both pontocerebellar corners to the internal acoustic canal, hyper-intensity in T2-concentrated sequences, and massive lesions with regular, lobulated contours heavily retaining contrast following injection of the contrast medium were observed. Massive lesions were causing significant pressure on the brain stem and the fourth ventricle, causing obstructive hydrocephalus (Figure 2). In addition, contrast-retaining intradural intra-extramedullary masses, as well as multiple neurofibromas extending from neural foramina into the parasagittal soft tissues were noted in the spinal MRI scan (Figure 3).

The patient was admitted to an emergency surgery ward for treatment of hydrocephalus, and a ventriculoperitoneal shunt was

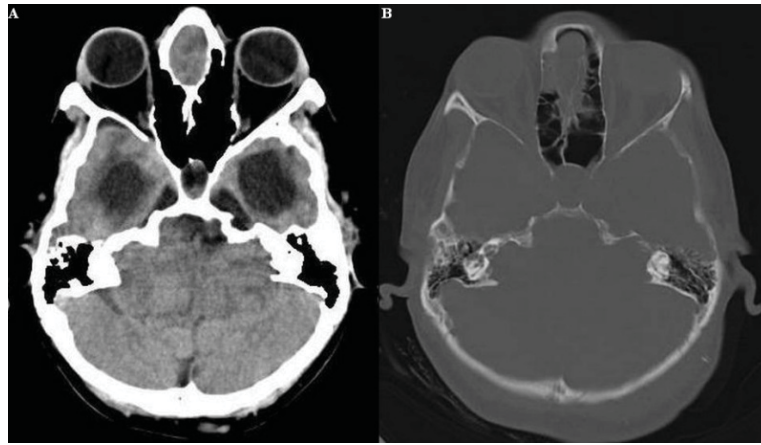


Figure 1. In the brain CT scan; **A)** hypo- isodense massive lesions with vague contours are observed in the parenchymal window in both pontocerebellar corners, and lateral ventricle temporal horns appear significantly dilated; **B)** both internal acoustic canals are observed to have been significantly widened by the component of the tumor inside the canal and have partially eroded contours in the bony window.

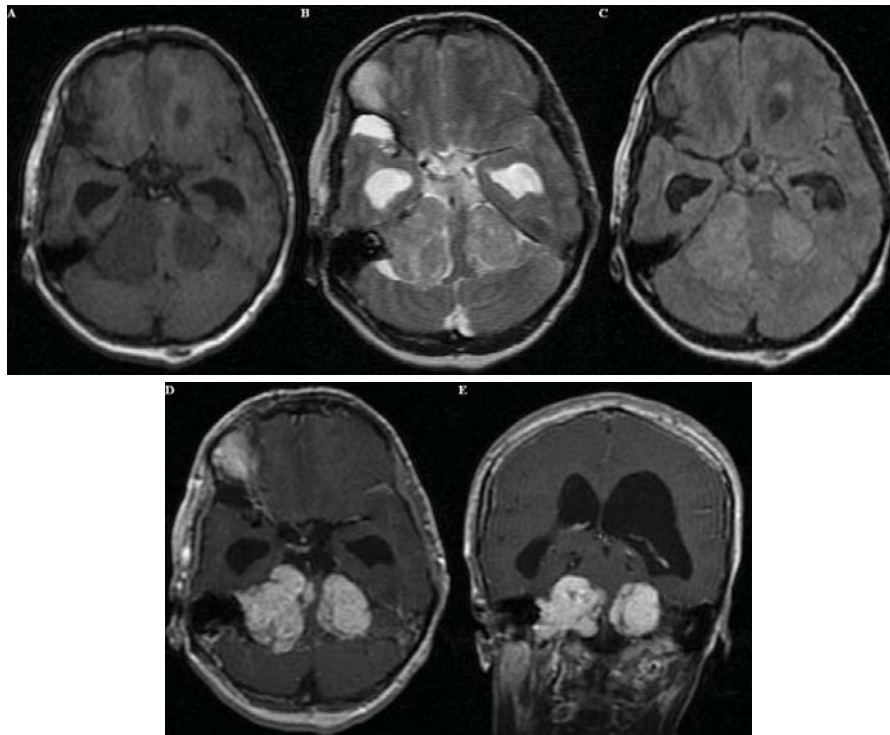


Figure 2. In the brain MRI scan, bilateral vestibulocochlear schwannomas extending into the internal acoustic internal canal in both pontocerebellar corners in the axial T1; **A)** axial T2; **B)** axial Flair; **C)** and axial contrasted T1; **D)** and coronal contrasted T1; **E)** images are observed. The masses constitute significant pressure on the brain stem and the fourth ventricle and cause obstructive hydrocephalus.

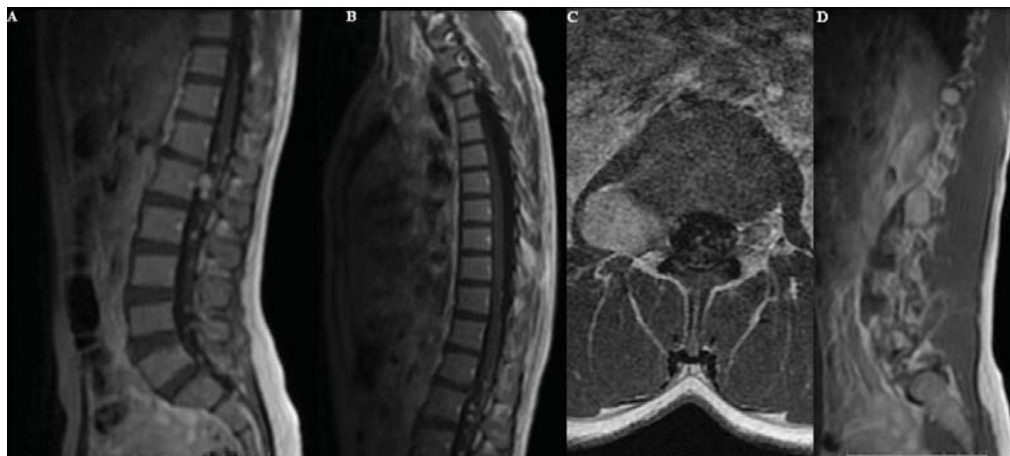


Figure 3. In the spine MRI scan, intradural intra-extramedullary masses; **A, B)** retaining contrast medium in the thoraco-lumbar spinal canal, and neurofibromas; **C, D)** composed of neural foramen and extending into the parasagittal soft tissue are observed.

inserted. The patient was taken into post-operative intensive care. Her consciousness did not improve. The patient was followed up at the intensive care unit for one week in an intubated fashion; her exitus took place as a result of cardiac arrest. The reason for exitus of the patient was considered to be cerebral perfusion disorder that developed as secondary to hydrocephalus.

Discussion

Vestibular schwannoma, which constitutes the majority of cerebellopontine angle tumors, is observed in two different groups of patients in the population. Unilateral vestibular schwannoma is encountered in elderly patients and sporadically, whereas bilateral vestibulocochlear schwannoma appears in NF-2 patients.²

National Institute of Health consensus committee has determined that bilateral masses of the eighth cranial nerve are diagnostic for NF-2. This disease is also called the syndrome of multiple inherited schwannomas, meningiomas, and ependymomas (MISME).³

Schwannomas of other cranial nerves develop more frequently in NF-2, and the presence of one of the rare cranial nerve schwannomas is generally an indication of the possibility of NF-2.

Several authors have studied a series of cases and tried to determine the incidence of tumors in cases of NF-2. Mautner, et al. studied 48 patients with NF-2 and found vestibular schwannomas in 46 patients, spinal tumors in 43, posterior sub-capsular cataracts in 30, meningiomas in 28, and trigeminal schwannomas in 14.⁴ Aoki, et al. reported on the cranial MRI scans of 11 patients. In their series, all patients had vestibular schwannomas, 8 had other cranial nerve tumors, and 6 had meningiomas.⁵ Patronas, et al. studied a series of 49 patients with NF-2 using spinal MRI images, which revealed spinal cord and/or canal tumors in 31 patients. Twenty-six patients had intramedullary lesions, 27 patients had intradural extramedullary tumors, and 22 patients had at least one tumor of each type.⁶

Our case was observed to have bilateral giant vestibulocochlear schwannomas. We detected a few small, enhancing intramedullary lesions in the distal thoracic spinal cord of our patient. We initially suspected by the diagnoses of ependymoma or astrocytoma. Additionally, multiple intradural extramedullary masses (meningioma/neurofibroma) along the entire spinal cord, and multiple neurofibromas causing the appearance of dumbbell-shaped tumors of different sizes in the neural foramina were observed. The patient had bilateral hearing loss; her ocular findings were normal.

There are a number of reports of patients who have been presented with hydrocephalus secondary to NF-1. The underlying pathologies of these cases were as follows: periaqueductal/tectal hamartomas, hamartomas in the cerebellum-tegmentum, brain stem glioma, mesencephalic pilocytic astrocytoma, synechia, aqueductal web, periaqueductal gliosis, and stenosis.⁷ Similarly, there are reports in the literature of cases presenting with hydrocephalus secondary to vestibulocochlear schwannoma.⁸ In our case, hydrocephalus developed in an NF-2 patient. The most important aspect of our case was that vestibulocochlear schwannomas caused hydrocephalus that led to death for the first time.

Bilateral giant vestibulocochlear schwannomas caused pressure on the brain stem and the 4th ventricle, resulting in hydrocephalus, which later led to the patient's death. To the best of our knowledge, this is the first case where bilateral vestibulocochlear schwannoma with NF-2 has resulted in hydrocephalus.

In conclusion, bilateral vestibulocochlear schwannoma may cause hydrocephalus in NF-2. Although it is rare, bilateral vestibulocochlear schwannoma may lead to death if it is not treated. Therefore, it is recommended to carefully monitor and treat bilateral vestibulocochlear schwannoma in accordance with its stage.

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