Case Report

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MRI Evaluation of Neurofibromatosis Type 2 in a Young Female with Vestibular Schwannoma and Multiple Meningiomas: A Case Report

Radiology Section

SUVANSH BHATIA[1], KUNAL SOLANKI[2], MAULIK MUKESHBHAI KOTHARI[3], OJUS SETH[4], SAMARTH SHAH[5]

ABSTRACT

Neurofibromatosis Type 2 (NF2) is a rare autosomal dominant neurocutaneous disorder characterised by the development of multiple central nervous system tumours, particularly vestibular schwannomas and meningiomas. Magnetic Resonance Imaging (MRI) plays a crucial role in early detection and surveillance of these lesions. A 22-year-old female presented with progressive hearing loss in the right ear for five months and intermittent headache. Pure tone audiometry revealed moderately severe sensorineural hearing loss in the right ear. MRI brain demonstrated a right cerebellopontine angle extra-axial mass extending into the internal auditory canal. The lesion appeared T2/FLAIR hyperintense, showed no diffusion restriction, demonstrated few blooming foci on susceptibility weighted imaging, and showed homogeneous postcontrast enhancement. The patient subsequently underwent surgical excision of the lesion, and histopathological examination of the excised specimen confirmed schwannoma. Additionally, two lesions consistent with meningiomas were identified. The first was an extra-axial lesion in the left parietal region which appeared heterogeneously hyperintense on T2/FLAIR. The second lesion was a calcified lesion located in the temporal horn of the right lateral ventricle, appearing hypointense on both T1 and T2/FLAIR sequences and demonstrating blooming on susceptibility-weighted imaging without diffusion restriction. The co-existence of vestibular schwannoma with multiple intracranial meningiomas raises suspicion for neurofibromatosis Type 2. MRI plays a key role in detection and follow-up of patients with suspected NF2.

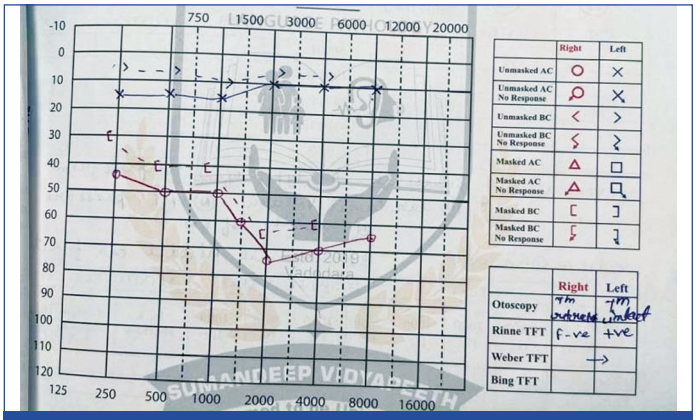
Keywords: Cerebellopontine angle tumour, Intraventricular meningioma, Magnetic resonance imaging, Neurocutaneous syndrome, Sensorineural hearing loss

CASE REPORT

A 22-year-old female presented with progressive hearing loss in the right ear for five months associated with intermittent headache. A few non specific hyperpigmented macules were noted over the upper back. No cutaneous neurofibromas or characteristic cutaneous manifestations of NF2 were identified. There was no history of seizures, vomiting, trauma or visual disturbance. No family history of hearing loss, intracranial tumours or neurocutaneous disorders was reported. However, detailed pedigree analysis and genetic testing could not be performed in the present case.

On examination, the Romberg test was positive to the right, and the patient had an impaired gait (tandem gait). Cranial nerve examination revealed right-sided sensorineural hearing loss without evidence of facial nerve palsy, trigeminal nerve involvement, or lower cranial nerve deficits. Motor and sensory examination of all four limbs was unremarkable. Deep tendon reflexes were preserved and plantar responses were flexor bilaterally. Cutaneous examination revealed a few small hyperpigmented macules over the upper back. No cutaneous neurofibromas were identified. Ophthalmological examination was unremarkable.

Pure tone audiometry revealed moderately severe sensorineural hearing loss in the right ear [Table/Fig-1]. MRI of the brain was performed for further evaluation. MRI revealed a well-defined extra-axial lesion in the right cerebellopontine angle measuring 2.7×2.5×3.1 cm (AP×TR×CC) extending into the right internal auditory canal, with widening of the canal. The lesion appeared hyperintense on T2/FLAIR sequences and showed no diffusion restriction. A few blooming foci were noted on susceptibility-weighted imaging. Post-contrast T1-weighted images demonstrated homogeneous enhancement. The lesion caused mass effect in the form of compression and medial displacement of the right cerebellar peduncle [Table/Fig-2]. Imaging findings were suggestive of vestibular schwannoma.

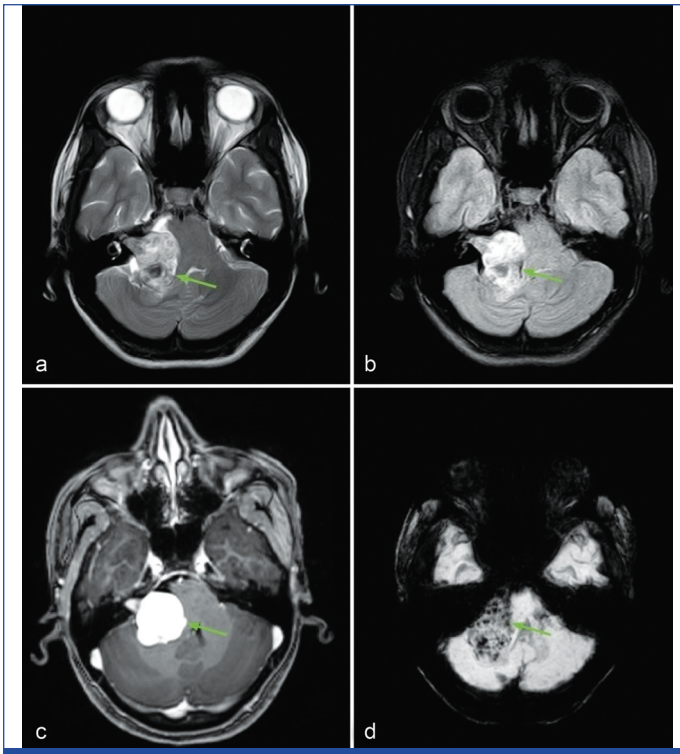


[Table/Fig-1]: Pure tone audiometry demonstrating moderately severe sensorineural hearing loss in the right ear.

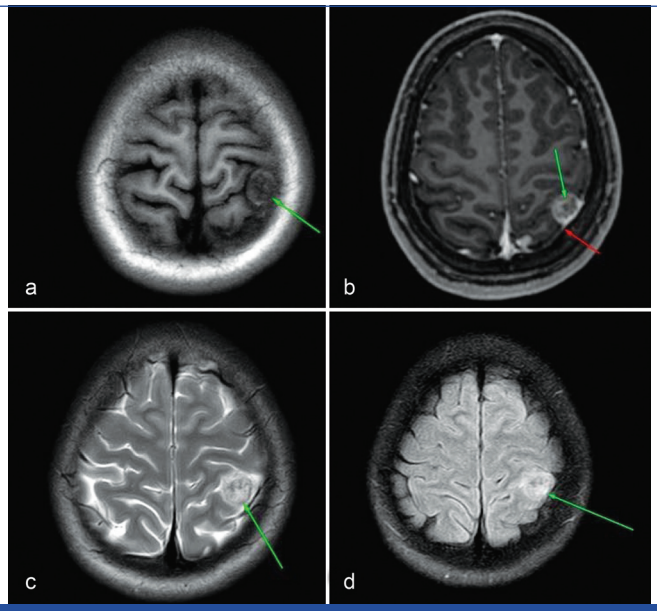
In addition, two lesions consistent with meningiomas were detected. The first lesion was an extra-axial lesion located in the left parietal region measuring 1.5×1.5×1.2 cm. It appeared heterogeneously hyperintense on T2/FLAIR and hypointense on T1-weighted imaging with heterogeneous post-contrast enhancement and showing a dural tail sign. No blooming foci or diffusion restriction were observed [Table/Fig-3].

The second lesion was in the temporal horn of the right lateral ventricle measuring 1.2×0.9×0.8 cm. The lesion appeared hypointense on both T1 and T2/FLAIR sequences and demonstrated blooming on susceptibility weighted imaging consistent with calcification. No diffusion restriction was seen. Axial postcontrast MRI images showing a hypointense lesion with heterogenous postcontrast enhancement [Table/Fig-4].

The patient subsequently underwent surgical excision of the cerebellopontine angle tumour. Histopathological examination of the excised specimen confirmed the diagnosis of schwannoma [Table/Fig-5].



[Table/Fig-2]: MRI images demonstrating a right cerebellopontine angle mass (green arrows): a) Axial T2-weighted MRI showing a well-defined hyperintense extra-axial lesion in the right cerebellopontine angle extending into the internal auditory canal; b) Axial FLAIR image demonstrating hyperintense signal within the lesion; c) Postcontrast T1-weighted MRI showing homogeneous enhancement of the lesion; d) Susceptibility-weighted imaging demonstrating few blooming foci within the lesion, consistent with vestibular schwannoma.



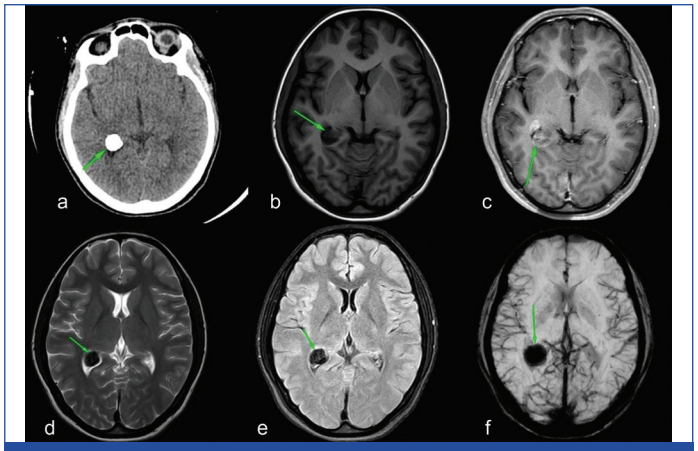
[Table/Fig-3]: a) Axial T1-weighted MRI showing a hypointense extra-axial lesion along the left parietal convexity (green arrow); b) Postcontrast T1-weighted MRI showing heterogeneous enhancement of the lesion (green arrow) and showing dural tail sign (red arrow); c,d) Axial T2/FLAIR image demonstrating heterogeneous hyperintensity within the lesion (green arrows). Findings suggestive of meningioma.

The imaging and histopathological findings raised a strong clinical suspicion of Neurofibromatosis Type 2; however, genetic confirmation was not available.

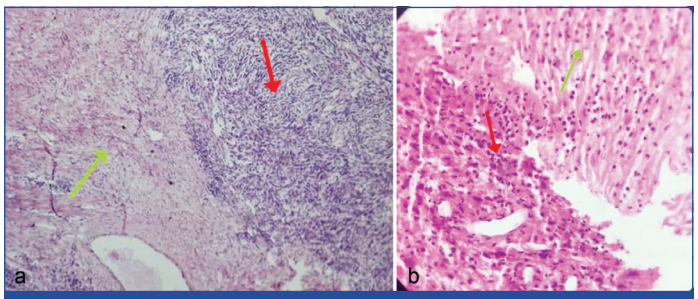
Following surgery, the patient was referred for multidisciplinary evaluation and advised regular clinical and radiological follow-up. MRI evaluation of the spine was also recommended as part of ongoing surveillance.

DISCUSSION

Neurofibromatosis Type 2 (NF2) is a rare autosomal dominant neurocutaneous disorder caused by mutations of the NF2 tumour suppressor gene located on chromosome 22, which encodes



[Table/Fig-4]: CT and MRI images demonstrating a lesion within the temporal horn of the right lateral ventricle (green arrows): a) Non-contrast CT image showing a hyperdense calcified lesion in the temporal horn of the right lateral ventricle; b) Axial T1 weighted; and (c) Axial Post contrast MRI images showing a hypointense lesion with heterogenous post-contrast enhancement; d and e) Axial T2 and FLAIR MRI images demonstrating corresponding hypointense signal; f) Susceptibility-weighted imaging demonstrating prominent blooming consistent with calcification, suggestive of calcified intraventricular meningioma.



[Table/Fig-5]: Histopathological examination (H&E stain) at 4× (a) and 40× (b) magnification demonstrating a biphasic tumour composed of compact hypercellular Antoni A areas (green arrows) and loose hypocellular Antoni B areas (red arrows). Spindle-shaped tumour cells are identified within the Antoni A region.

the protein merlin [1,2]. The disorder is characterised by the development of multiple nervous system tumours, most commonly vestibular schwannomas, meningiomas and ependymomas [1,2]. NF2 is uncommon, with an estimated incidence of approximately 1 in 25,000-40,000 individuals [1,2]. Vestibular schwannomas are the hallmark lesions of NF2 and typically present with progressive sensorineural hearing loss, tinnitus and balance disturbances [1,2]. The co-existence of a histopathologically confirmed vestibular schwannoma with multiple intracranial lesions consistent with meningiomas raises a strong clinical suspicion of NF2-related schwannomatosis. According to the Manchester diagnostic criteria, the presence of vestibular schwannoma in association with multiple intracranial tumours strongly supports the clinical diagnosis even in the absence of a positive family history [3].

Available treatment options include observation with serial imaging, microsurgical excision and stereotactic radiosurgery [4]. Preservation of hearing remains an important therapeutic goal, particularly in young patients, and treatment decisions should balance tumour control with maintenance of neurological function and quality of life [4,5].

The differential diagnosis of a cerebellopontine angle mass includes meningioma, epidermoid cyst, arachnoid cyst, metastatic disease and other cranial nerve schwannomas. However, extension into the internal auditory canal, widening of the canal, T2/FLAIR hyperintensity and homogeneous postcontrast enhancement strongly favoured vestibular schwannoma in the present case. Similarly, the intraventricular lesion was differentiated from choroid plexus papilloma, central neurocytoma and ependymoma based on its imaging characteristics, calcification and association with additional intracranial lesions. Histopathological confirmation of schwannoma further supported the diagnosis.

Recent case reports have described similar manifestations of NF2-related schwannomatosis. Hamza L et al., reported a patient with

MISME syndrome presenting with multiple intracranial schwannomas, meningiomas and ependymomas, emphasising the importance of comprehensive neuroimaging in detecting the full tumour burden [5]. In contrast, index patient demonstrated a unilateral vestibular schwannoma associated with two intracranial lesions consistent with meningiomas, without evidence of ependymoma. Similarly, Soni S and Yadav MS described classical MRI findings of NF2 with multiple intracranial tumours and characteristic imaging features [6]. However, index case was notable because the diagnosis was suspected primarily on the basis of a vestibular schwannoma associated with multiple meningiomas in the absence of genetic confirmation and a positive family history. These reports, together with the present case, highlight the pivotal role of MRI in raising suspicion for NF2-related schwannomatosis and guiding further clinical evaluation.

The present case highlights the importance of carefully searching for additional intracranial lesions in patients presenting with vestibular schwannoma. Recognition of multiple meningiomas in association with a vestibular schwannoma may provide an important clue to an underlying diagnosis of Neurofibromatosis Type 2, particularly in patients without a positive family history or genetic confirmation.

CONCLUSION(S)

The MRI played a pivotal role by demonstrating a cerebellopontine angle schwannoma with internal auditory canal extension along with multiple intracranial meningiomas, providing important imaging clues toward the diagnosis. Histopathological confirmation of schwannoma further strengthened the diagnostic impression.

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PARTICULARS OF CONTRIBUTORS:
1. Junior Resident, Department of Radiology, Smt. B.K. Shah Medical Institute and Research Centre, Sumandeep Vidyapeeth, Vadodara, Gujarat, India.
2. Professor, Department of Radiology, Smt. B.K. Shah Medical Institute and Research Centre, Sumandeep Vidyapeeth, Vadodara, Gujarat, India.
3. Junior Resident, Department of Radiology, Smt. B.K. Shah Medical Institute and Research Centre, Sumandeep Vidyapeeth, Vadodara, Gujarat, India.
4. Junior Resident, Department of Radiology, Smt. B.K. Shah Medical Institute and Research Centre, Sumandeep Vidyapeeth, Vadodara, Gujarat, India.
5. Resident, Department of Radiology, Smt. B.K. Shah Medical Institute and Research Centre, Vadodara, Gujarat, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Suvansh Bhatia,
Gargi Hostel, Vadodara, Gujarat, India.
E-mail: suvansh15@gmail.com

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