Neurofibromatosis type 2 (NF2) is a rare autosomal-dominant disorder with an incidence of approximately 1:40,000 to 1:50,000 individuals, and an equal gender predisposition. Although a disease of adults, it could be evident in childhood and often gets unrecognized. The disease has variable expression ranging from characteristic right and left vestibular schwannomas (VS), skin tumors, ocular abnormalities, and other cranial and spinal tumors. Some have nonneoplastic lesions such as schwannosis, meningioangiomatosis, and glial hamartia. Prognosis depends on the grade of severity, and as the disease is progressive, patient should remain under surveillance. The affected individuals should be advised not to have children.

Case Report

A 17-year-old male patient, product of non-consanguineous marriage, presented with complaints of weakness and wasting of left leg for 5 years, reduced hearing in both ears for 6 months, and weakness and wasting of left hand for 4 months. He had multiple nodules over the trunk, right knee, and left suboccipital region. There was no evidence of clinical manifestations of neurofibromatosis in his family members, when traced out up to 3 generations. There was no history of associated headache, fever, seizures, vomiting, unconsciousness, ataxia, vertigo, difficulty in vision, facial deviation, giddiness, nasal regurgitation of food, difficulty in speech, memory loss, neck pain, and urinary/fecal incontinence. On examination, the nodules were found to be non-tender, firm, well defined, irregular, and of variable sizes. There was also a suboccipital and postauricular swelling on the left side with no cough impulse. He had bilateral asymmetric sensorineural hearing loss, and left foot drop and pes cavus. On ophthalmological examination, the patient was found to have a visual acuity of 6/9 and 6/18, which did not improve with pinhole. He had manifest squint with no restriction of eye movements or diplopia or nystagmus. He had wasting in his left lower limb and left hand. He had generalized loss of deep tendon reflexes with preservation of superficial reflexes. Fasciculations were seen in both the calves. The patient also had sensory loss to touch/pain in L5 dermatomal region. However, joint position and vibration sense were intact. There were no signs of incoordination or any autonomic involvement. On ultrasonography of the abdomen, a hypoechoic lesion was seen.

Keywords: Cranial nerves, MISME syndrome, neurofibromatosis

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measuring 11 × 12 mm was seen in pelvis indenting on the right wall of urinary bladder. On nerve conduction studies, demyelinating pattern was observed. Pure-tone audiometry confirmed bilateral sensorineural hearing, which was profound on the right and moderate on the left. Brainstem evoked response audiometry of the patient revealed no response on the right and only one wave on the left.

Review of the contrast enhanced MR imaging of brain showed large multilobulated solid-cum-cystic lesion in cerebellar verminian region, which continued posteriorly into retrocerebellar, suboccipital, and brain stem region. It exhibited enhancement of its solid mural nodular component, which was compressing on brain stem, craniovertebral, and cervico-medullary junction [Figure 1]. There was also inhomogeneous patchy enhancement of both cerebellopontine (CP) angle region and prepontine region along the course of vestibulocochlear and trigeminal nerves, in Meckel cave region and bilateral cavernous sinus regions. Both vestibulocochlear nerves in CP angle regions were involved with intracanalicular extension of lesions [Figure 2A]. Paracavernous region exhibited bilateral involvement of multiple cranial nerves namely 3rd, 4th, and 6th; extra-calvarial retrocerebellar, retro-occipital, and suboccipital regions also exhibited multiple nodular-enhancing lesions of variable sizes representing neurocutaneous nodules with a possibility of plexiform involvement [Figure 2B]. Hydrocephalus presented with dilated temporal horns. Both optic nerves exhibited patchy enhancement along their course in orbital apex and optic tracts.

The MRI of the spine showed diffuse patchy heterogeneous hyperintense signal in T2-weighted sagittal images exhibited as subtle patchy enhancement in cervical and conus regions. There were multiple enhancing extradural nodular lesions with broad base at C6-C7 and D5-D6 levels, extending through both lateral neural regions and C5-C6 levels, and occupying right lateral aspect of spinal canal at D5-D6 levels. There was perineural involvement of caudal filum [Figure 2C].

Biopsy of the cutaneous lesion was consistent with neurofibroma [Figure 3]. Although the patient had no family history, the image findings of bilateral acoustic schwannomas, multiple intracranial meningiomas, and intramedullary tumor suggested the diagnosis of NF2.

**Discussion**

We present a case of NF2 with bilateral multiple cranial nerve schwannomas involving 3rd, 4th, 5th, 6th, 8th, 9th, 10th, 11th, and 12th cranial nerves with early-onset polyneuropathy. Our case presents as left-sided weakness of both upper and lower limbs with bilateral hearing loss, which on workup was found to be a case of NF2 with bilateral multiple cranial nerve involvement. NF2 is a syndrome with bilateral VS, which is its pathognomonic and diagnostic criteria.[5-7] In its initial assessment, there are chances for these bilateral VS to be missed especially in patients without any family history. Of all the cases, 50% constitute first in family most probably because of the somatic mutation of NFT gene. Similar was the situation in our case, with no family history for the past three generations.

NF2 has a varied clinical presentation with majority being presented with hearing loss accompanied or preceded by tinnitus because of the involvement of the 8th cranial nerve, which is similar to our case.[8] Studies have reported weakness and wasting of the muscle groups similar to poliolike features, years before detection of VS. Cases have also documented polyneuropathy in adulthood.[8] Weakness of lower limb muscles at the age of 12 years and polyneuropathy, occurring only in 3%–5%, in the form of involvement of hand muscles before adulthood could suggest the aggressiveness of the disease.
The average age of onset of clinicoradiological findings has been found to be 18–24 years and in the range of birth to 70 years.[9] Being labeled as an adult-onset disease, it could get under-recognized in children.[9] This happened in our case too, when the patient developed first symptom in the form of lower limb weakness at the age of 12 years without any skin lesion or visual complaints.[9] Age of symptom onset is a very crucial feature, defining the prognosis of the disease. Early onset of initial symptoms has been reported to significantly decrease the survival rate.[10]

Literature suggests that schwannomas can involve any of the cranial nerve except optical nerve.[8] We present this as a case of NF2 with extensive involvement (3rd, 4th, 5th, 6th, 8th, 9th, 10th, 11th, and 12th) of cranial nerves along with diffuse spinal involvement. The last reported case was by Pasricha et al.,[11] in which they had diagnosed lesions affecting seven cranial nerves in NF2. To the best of our knowledge, no other paper has reported such a case that involves tumors affecting nearly all cranial nerves (namely 3rd–12th nerves) with diffuse spinal involvement, their association with meningiomas and intramedullary spinal cord tumors, optic lesion, and early-onset polynuropathy in the same patient.[11]

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Conflicts of interest
There are no conflicts of interest.

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