Case Report

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Bilateral vestibular schwannoma with a cooccurring meningioma in a child: a case report and review of literature

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Introduction: Meningioma and vestibular schwannoma (VS) are the first and second most common benign central nervous system tumors. The coexistence of VS and meningioma presents a rare clinical scenario, particularly in pediatric patients. This report presents a case of bilateral VS with a cooccurring meningioma in a Nepali child and provides an overview of the literature on this condition.

Case report: A 15-year-old male presented with bilateral sensorineural hearing loss, seizures, and neurological deficits and was ultimately diagnosed with concomitant bilateral acoustic neuroma and meningioma. The patient underwent radiosurgery for bilateral VS and nonoperative management of the meningioma. Long-term follow-up revealed symptomatic improvement, emphasizing the importance of a multidisciplinary approach in managing such complex cases. The management of these tumors requires tailored treatment strategies guided by tumor characteristics and associated risks.

Discussion: Meningioma and VS are common tumors of the central nervous system. Their coexistence is possible in neurofibromatosis type 2 but is exceedingly rare in pediatric age group. The tumors, often coexisting, pose diagnostic challenges. Diagnosis relies on clinical and genetic features, with multidisciplinary management involving various specialists. Treatment aims to preserve function and quality of life, utilizing approaches such as bevacizumab and surgical intervention. The role of radiation therapy remains uncertain. Genetic testing and regular monitoring are vital for early detection and intervention.

Conclusion: The cooccurrence of acoustic neuromas and meningiomas is poorly understood, with limited reported cases and unclear pathophysiological mechanisms. Further research into the genetic and molecular mechanisms underlying the coexistence of these tumors is needed to optimize patient outcomes in this rare clinical entity.

Keywords: case report, meningioma, Nepal, neurofibromatois type 2, vestibular schwannoma

Introduction

VS is a benign intracranial tumor that originates from schwann cells that reside along the vestibular portion of the eighth cranial nerve[1]. VS accounts for 10% of all CNS tumors and 80% of the tumors in the cerebellopontine angle[2,3]. Meningioma derives from the meningothelial cells of the arachnoid layer and is the most common benign CNS tumor, accounting for 13–26% of all brain tumors overall. More than 90% of all meningiomas are solitary[1]. However, in some extremely rare situations, VS can coexist with a meningioma in an adjacent anatomical position. This situation is most often reported in patients with NF2 or those who have a history of previous irradiation[4–7]. NF2 is an autosomal dominantly inherited syndrome that predisposes individuals to bilateral VSs as well as multiple other tumors of the nervous system[8–10]. Except for those circumstances, the coexistence of these tumors is exceedingly rare. Children diagnosed with NF2 often have an atypical but more severe presentation. Such a combination is exceptionally rare in this age group and emphasizes some unique aspects of diagnosis and management.

HIGHLIGHTS

- Meningioma and vestibular schwannoma (VS) are the first and second most common benign central nervous system (CNS) tumors.
- Their coexistence is possible in neurofibromatosis type 2 (NF2) but is exceedingly rare in the pediatric age group.
- Diagnosis relies on clinical and genetic features, with multidisciplinary management involving various specialists.
- Further research into the genetic and molecular mechanisms underlying the coexistence of these tumors is needed to optimize patient outcomes in this rare clinical entity.

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Herein, we present a rare case of a 15-year-old male with concomitant bilateral VS and meningioma, which is also the first such reported case from Nepal, and a review of literature of similar other cases. This is reported in line with the Surgical CAse REport (SCARE) 2023 criteria[11].

Case report

History

A 15-year-old male presented to the outpatient department with the complaint of bilateral hearing loss, occasional imbalance, and several episodes of generalized tonic–clonic seizures. The hearing loss initially started almost 2 years back from the left ear and progressed to involve both ears. There were multiple episodes of tinnitus during the progression of hearing loss. He also complained of blurred and double vision and early morning headaches. He had several episodes of generalized tonic–clonic seizures in the last 2 months, which led his parents to seek medical care. He was not treated in any other center for such complaints in the past. There was no known family history of NF2. There was no history of skull irradiation in the past.

Examination

On examination, he was well oriented to time, place, and person, had a Glasgow Coma Scale of 15 (E4M5V6), had normal vital signs, and there was no pallor, icterus, clubbing, cyanosis, or edema. Neurological examination revealed bilateral sensorineural hearing loss (positive Rinne test and no lateralization of Weber test), papilledema, nystagmus, diplopia on lateral gaze, decreased corneal reflexes, facial twitching, and hyperesthesia. Other cranial nerves, motor and sensory systems, and cerebellar examination were normal. Physical examination did not reveal any findings suggestive of NF2.

Investigations

Preoperative MRI of the brain revealed the following findings (Fig. 1):

- Well-defined multilobulated extra-axial lesions of altered signal intensity (hypointense in T1, heterogeneously isointense to gray matter) in bilateral cerebellopontine angle cisterns extending into the internal acoustic meatus measuring 30×25 mm and 28×17 mm in left (thin white arrow) and right side (broad white arrow), respectively, were present. The lesions were abutting both sides of the pons and anteromedial aspect of bilateral cerebellar hemispheres. Similar lesions measuring 17×14 mm and 13×11 mm were extending into bilateral Meckel's cave. The lesions showed mild homogenous enhancement after contrast. There was no evidence of restriction of diffusion. These lesions were suggestive of bilateral vestibular and trigeminal schwannomas.
- A well-defined intraventricular lesion of altered signal intensity (isointense to gray matter in both T1 and T2) measuring 22×21 mm is noted in the posterior horn of the right lateral ventricle, and expanding it with restriction of diffusion was present (white arrowhead). Mild edema in the adjacent brain parenchyma was also noted. In the postcontrast study, the lesion showed near-homogenous and moderate enhancement. There was no evidence of intralesional necrosis, hemorrhage, or calcification. These features were suggestive of a meningioma (differential diagnosis: choroid plexus papilloma).

Pure-tone audiometry demonstrated bilateral severe sensorineural hearing loss. MRI of the spine was normal. Other laboratory investigations, including complete blood count, renal and liver function tests, and serology, were normal.

Treatment and follow-up

The patient's parents were counseled regarding the possible diagnosis of NF2. Subsequently, they expressed interest in seeking CyberKnife Radiosurgery in India. It was chosen due to the risk of neurological deficits after surgical resection and other surgical risks to the patient. An informal follow-up via phone call revealed that the patient underwent radiosurgery for bilateral VSs. Following a comprehensive assessment of the meningioma's characteristics and location, a nonsurgical approach was selected due to the potential risks and absence of acute symptoms. This decision was reached through collaborative efforts involving neurosurgeons, neurologists, and neuroradiologists. Following surgical intervention for the VS and conservative management of the concurrent meningioma, the patient reported improvement in symptoms, particularly in hearing and balance. Any kind of genetic testing was not done due to limited access and financial reasons. A follow-up MRI of the brain conducted 32 months later revealed the following findings (Fig. 2):

- Well-defined altered signal intensity lesion (T1 hypointense and T2/FLAIR heterogeneously hyperintense) of size 22×20 mm in the atrium of right lateral ventricle with homogenous enhancement on postcontrast study and areas of diffusion restriction on DWI within the lesion likely an intraventricular meningioma (white arrowhead).
- Well-defined homogenously enhancing multilobulated T1 hypointense and T2/FLAIR isointense lesions of size 24×23 mm and 25×12 mm were present in the right and left cerebellopontine angles, respectively, with evidence of extension into the internal auditory angle and prepontine cistern and homogenous enhancement postcontrast likely bilateral VSs (broad and thin white arrows respectively).
- Two well-defined homogenously lobulated altered signal intensity lesions measuring 18×16 mm and 19×10 mm in the right and left prepontine cisterns, extending into adjacent Meckel's cave and heterogenous enhancement postcontrast likely bilateral trigeminal schwannomas.
- T2 hypointense lesion of size 26×8.3 mm arising from the posterior duramater at D6–D7 level likely a spinal meningioma (differential diagnosis: spinal schwannoma) with compression of the adjacent spinal cord with T2 hyperintensity.
- Multiple T2 hypointense nodular lesions arising from the cauda equina and filum terminale are likely to be neurofibromas

Discussion

Meningioma and VS are the first and second most common benign CNS tumors. The coexistence of these tumors is seen relatively often in patients with NF2, which is an autosomal dominantly inherited syndrome that predisposes individuals to bilateral VSs as well as multiple other tumors of the nervous system[8–10]. A previous history of irradiation is another known cause of the occurrence of multiple primary brain tumors[12].

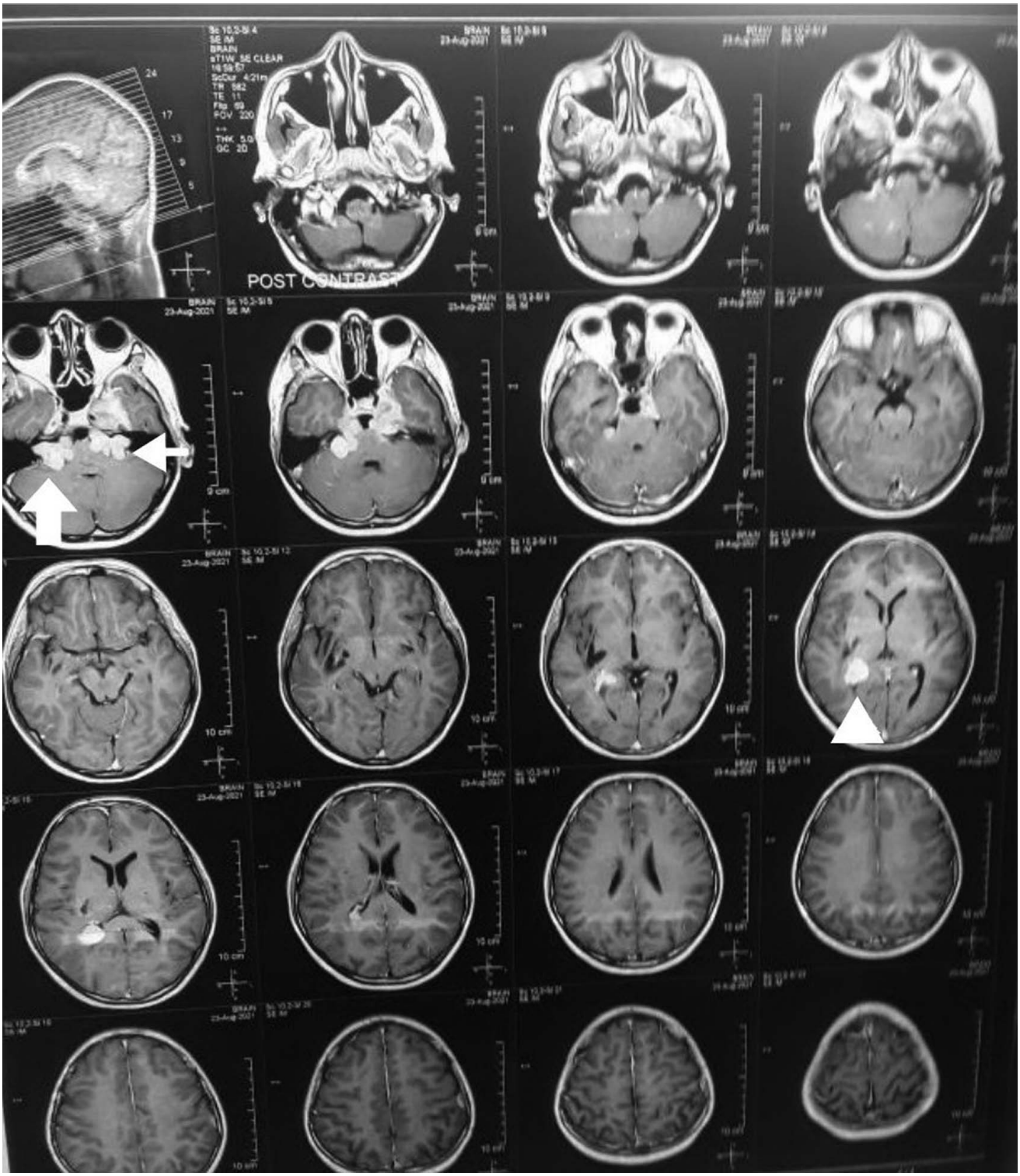

Figure 1. Preoperative MRI of the brain.

Generally, acoustic neuromas are diagnosed between the fourth and sixth decades of life. However, individuals with NF2 tend to present earlier, with the peak incidence occurring in the third decade of life[13]. Children diagnosed with NF2 often have an atypical but more severe presentation[14]. In childhood, patients commonly present with a constellation of symptoms, including eye problems, hearing loss, weakness, pain, mononeuropathy, cutaneous tumors, and seizures[15–17]. In contrast, adults with NF2 commonly present with hearing loss and tinnitus as the predominant symptoms. The hallmark features of NF2

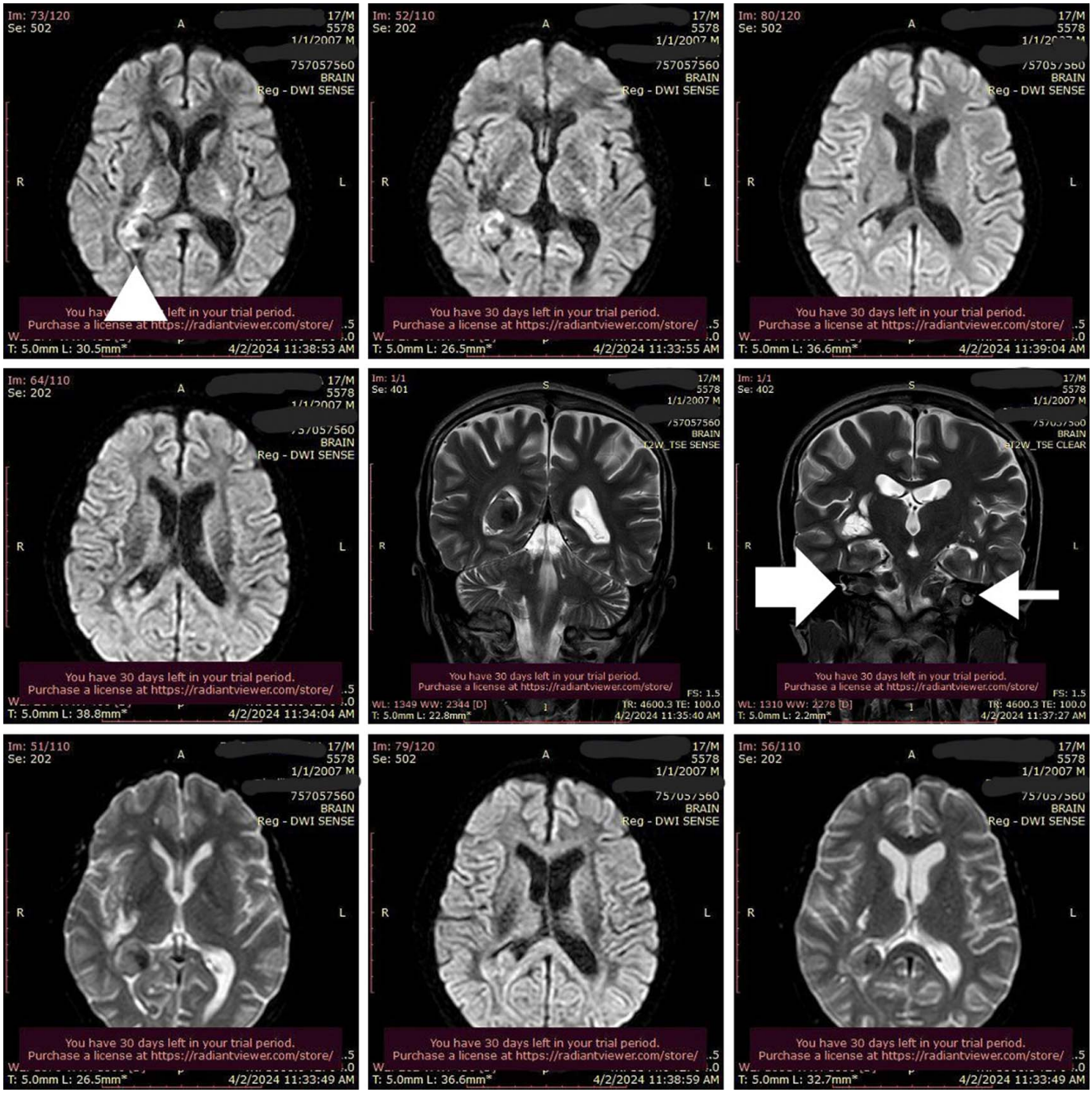

Figure 2. Postoperative MRI of the brain.

encompass a spectrum of neurologic and ocular lesions, including bilateral VSs, schwannomas of other cranial nerves, intracranial meningiomas, intramedullary and extramedullary spinal tumors, and peripheral neuropathy[18–20]. Additionally, ocular manifestations such as cortical and posterior subcapsular cataracts, epiretinal membranes, and retinal hamartomas are frequently observed[21–23]. Cutaneous manifestations, including cutaneous tumors, skin plaques, and subcutaneous tumors, further contribute to the clinical complexity of NF2[18,20,24,25].

VSs, the primary hallmark lesion of NF2, are typically bilateral and cause symptoms such as tinnitus, hearing loss, and balance dysfunction[10]. Hearing loss is well-documented as the most common presenting symptom in patients with VSs[26–34]. The onset of hearing loss is typically gradual and progressive, leading to deafness over time, although sudden hearing loss can also occur. NF2-related VSs are usually multifocal, affecting both the superior and inferior vestibular nerves[35]. Despite their prevalence, the mechanism underlying hearing loss in NF2-related VSs remains poorly understood, with no clear correlation between tumor size, growth rate, and the degree of hearing loss observed[36–39]. If left untreated, VSs can extend medially and cause brainstem compression and hydrocephalus.

Individuals with NF2 commonly develop meningiomas, with about half of NF2 patients affected[40]. The incidence of

meningiomas increases with age, potentially reaching a lifetime risk of up to 75%[20]. While most meningiomas are found within the cranium, spinal meningiomas are also observed. NF2 patients typically develop meningiomas at a younger age compared to sporadic cases, with approximately 20% of childhood meningioma diagnoses being associated with NF2[14,41]. Histopathologically, NF2-associated meningiomas are more frequently atypical or anaplastic compared to sporadic cases[42,43].

When meningiomas and VSs occur simultaneously, they are termed concomitant, collision[44], concurrent[45,46], coexisting[45–47], or coincidental tumors[2]. Several hypotheses have been proposed to explain the formation of these collision tumors:

1. Schwannomas may influence the local microenvironment, promoting the occurrence of meningiomas[5,48–50].
2. Mutations in the Merlin gene, located on chromosome 22q, are associated with meningioma development[51]. The absence or inactivation of merlin is also implicated in sporadic VSs[52].
3. Both neoplasms may originate from the same mesenchymal progenitor cells, differentiating into distinct tumor cell types[48,53–57].
4. Exposure of two different tumor cell types to the same oncogenic stimulus at the same site may lead to the development of a collision tumor[2,55,56].
5. The independent development of both neoplasms, with their concurrent appearance, is considered a coincidence[2,55,58].

The diagnosis of NF2 relies on characteristic clinical and molecular genetic features. The diagnostic criteria are recommended by an international consensus group[59], a refinement of the Manchester criteria. Diagnosis involves a comprehensive evaluation, including clinical and family history, neurological examination, and imaging studies such as contrast-enhanced MRI of the brain and spine[8]. MRI with gadolinium contrast is crucial for assessing meningiomas and VSs[45], although it may not always detect coexisting tumor types[45–47,60,61]. VSs often invade the internal auditory canal, exhibit cystic changes, and show reduced internal homogeneity on imaging[2]. In contrast, meningiomas typically present with a dural tail sign and calcifications[53,62], lacking in VSs. Imaging characteristics include intense to slight hypointensity on T1-weighted images and higher intensity on T2-weighted images for VSs compared to meningiomas[2,53,62,63]. Schwannomas are heterogeneously enhanced on contrast enhancement and frequently extend into the internal auditory canal, while meningiomas show homogenous enhancement[2,58]. Pathological examination reveals that schwannomas express S-100 protein and anti-Leu 7 but are negative for EMA, with possible focal EMA reactivity[5,46,64]. In contrast, meningiomas are positive for EMA and negative for S-100 and anti-Leu 7[2,5,64].

The management of NF2 is complex and requires a multidisciplinary approach involving various specialists to prevent or address potential complications. A multidisciplinary team typically includes neurosurgeons, neuro-otologists, neuroradiologists, neurologists or neuro-oncologists, audiologists, specialist nurses, and geneticists with expertise in NF2 care[65].

Treatment of VSs in NF2 patients aims at preserving function and maintaining quality of life[8]. Treatment is typically initiated when there is a risk of brainstem compression, hearing deterioration, or facial nerve dysfunction[10]. Bevacizumab, a monoclonal antibody targeting vascular endothelial growth factor, has shown promising results in inducing tumor shrinkage and improving hearing in NF2-associated VSs. Some centers use bevacizumab as a first-line therapy for rapidly growing schwannomas threatening function before considering surgery[66]. The efficacy of bevacizumab is supported by retrospective case series and limited prospective studies[66–76].

Surgical management is often necessary for VSs, although bevacizumab may be used initially in some cases. Surgical procedures in NF2 patients can be more complex due to multifocal tumors[35,77]. VSs may involve facial nerve fibers, increasing the risk of facial nerve damage during surgery[8].

The role of radiation therapy in NF2-related VSs is uncertain[78–81], with variable outcomes reported in studies. Long-term follow-up data are lacking, and concerns exist regarding the potential for increased risk of second malignancies after radiation therapy[82–84]. Additionally, surgical resection may be more challenging following stereotactic radiosurgery.

Hearing impairment in NF2 patients significantly impacts their quality of life and social well-being[85]. Strategies such as cochlear or brainstem implants may offer benefits for those with severe hearing impairment, helping to mitigate social avoidance, unemployment, and decreased social support associated with hearing loss in NF2[86].

Meningiomas can develop intracranially or within the spinal cord's intradural, extramedullary sites. Many NF2-associated meningiomas reach a stable size without needing active treatment. Factors like tumor size, peritumoral edema, absence of calcifications, and isointense or hyperintense MRI signals are risk factors for rapid growth, similar to sporadic meningiomas[87]. Rapidly growing or functionally threatening meningiomas are typically managed surgically, with radiation therapy considered for inoperable cases[88]. Lapatinib has shown some efficacy in progressive NF2-associated meningiomas[89], but evidence supporting bevacizumab's effectiveness is limited[90].

Intramedullary spinal tumors, primarily ependymomas, are often asymptomatic and grow slowly. Surgical resection is preferred over radiation therapy if intervention is necessary[91], although cystic components may respond to bevacizumab[92].

Genetic testing is vital for suspected schwannomatosis and NF2 diagnosis, especially in younger patients and first-degree relatives. Regular monitoring, including audiology, ophthalmologic evaluation, cutaneous examination, and MRI scans, is recommended for individuals with NF2 pathogenic variants[8,10,65,93]. The limitations of our study include lack of long-term follow-up data, lack of genetic testing of the patient, and for familial cases.

Conclusion

The cooccurrence of VSs and meningiomas is poorly understood, with limited reported cases and unclear pathophysiological mechanisms. While our study contributes to this knowledge gap, long-term follow-up data are needed, and genetic testing for familial causes remains unavailable. Further research into the genetic and molecular mechanisms underlying the coexistence of these tumors is needed to optimize patient outcomes in this rare clinical entity.

Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the patient's parents for the publication of this case report and accompanying images. A copy of written consent is available for review by the editor-in-chief of this journal on request.

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Author contribution

R.P.C.: wrote the original manuscript, reviewed and edited the original manuscript; K.K.C.: wrote the original manuscript, reviewed and edited the original manuscript; A.B.: wrote the original manuscript, reviewed and edited the original manuscript; Y.S.: edited the original manuscript.

Conflict of interest disclosure

There are no conflicts of interest.

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