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Original Article

Pediatric Intracochlear Schwannoma: Case Series and Review of the Literature

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BACKGROUND: Intracochlear schwannomas (ICSs) are a subtype of intralabyrinthine schwannomas, completely located in the cochlear lumen. ICSs are particularly rare in the pediatric population. Putative diagnosis is made on the basis of magnetic resonance findings with signal characteristics that should remain the same at follow-up imaging.

METHODS: A retrospective review was performed searching for pediatric patients affected by ICS treated at the Otolaryngology Department, Ospedale Ramazzini, Carpi (Italy), and Otolaryngology and Otoneurosurgery Department, Azienda Ospedaliero-Universitaria di Parma, (Italy). A scoping literature review of the period January 2000 - June 2024 was performed.

RESULTS: Two cases of ICS in pediatric patients are described. Neither family history nor genetic signs of neurofibromatosis type II were found. A single report was identified in the literature review. Data analysis resumes the pooled data of the latter case and the authors' patients. The most common symptom at presentation was progressive sensorineural hearing loss (66%). Mean pure tone average at diagnosis was 74.2 dB. Intracochlear location was in the basal turn in 2 cases and in the apical and middle turns in the third patient. All cases initially underwent a "wait and scan" strategy. The mean follow-up time was 23.3 months.

CONCLUSION: Management planning of pediatric ICSs should be accurate as surgical removal may require partial or total cochlear demolition, resulting in vestibular dysfunction and precluding future positioning of a cochlear implant. Close clinical and radiological follow-up with serial MRI scans allows to evaluate both symptom progression and rate of growth, in order to provide patients with the best therapeutic option.

KEYWORDS: Intracochlear schwannoma, pediatric schwannoma, sensorineural hearing loss, intralabyrinthine schwannoma

INTRODUCTION

Intralabyrinthine schwannomas (ILSs) are benign neoplasms arising within the boundaries of the inner ear.[1] Early symptoms include sudden or rapidly progressive hearing loss, dizziness, and tinnitus.[1,2] Magnetic resonance imaging (MRI) with gadolinium contrast is required for a definitive diagnosis.[3,4] Available therapeutic options include: "wait and scan" strategy, surgery, and radiotherapy.[2,5]

Intracochlear schwannomas (ICSs) are a subtype of ILS completely located in the cochlear lumen. ICSs are particularly rare in the pediatric population with only one case reported in the literature.[6]

In this report, two additional cases of ICS in pediatric patients are described: both complained of unilateral sensorineural hearing loss with no other audiovestibular symptoms. Neither family history nor genetic signs of neurofibromatosis type II (NF2) were found. A review of the published literature on the subject was performed covering the period January 2000 - June 2024.

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METHODS

A retrospective review was performed for pediatric patients affected by ICS treated at the Otolaryngology Department, Ospedale Ramazzini, Carpi (Italy), and Otolaryngology and Otoneurosurgery Department, Azienda Ospedaliero-Universitaria di Parma, (Italy) between May 2020 and June 2024. An evaluation of symptoms, results of audiological assessment, MRI findings, and management was performed. All patients underwent a complete head and neck examination, pure tone audiometry, speech discrimination, vestibular evaluation with the Head Shaking Test (HST), Head Impulse Test (HIT), and Unterberger test, a high-resolution MRI scan of the brain with gadolinium contrast, and CT scan of the temporal bone. Diagnosis of ICS was based on peculiar findings on high-resolution MRI scans: contrast enhancement of the cochlear lumen on T1-weighted images after gadolinium infusion, and sharply delineated signal intensity loss in the high-intensity intralabyrinthine fluids on T2-weighted images.

A scoping literature review of the period January 2000-June 2024 was also performed on PubMed and the EMBASE database by using the search terms "intralabyrinthine," "schwannoma," "pediatric," "acoustic," and "neurinoma" in different combinations. Abstracts on the subject were screened, and full texts were obtained when indicated.

This study did not need any approval by the local Institutional Review Board, as specifically indicated (version 4, 22.09.2020). The study was conducted in accordance with the principles of the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. All patients included in the study signed an informed consent. No animals were involved.

RESULTS

Over the period May 2020 - July 2024, two cases of ICSs in pediatric patients with no signs of neurofibromatosis type II were identified. Both patients were male. Both cases presented sensorineural hearing loss (SNHL) in the absence of tinnitus, dizziness, and facial weakness.

MAIN POINTS

- Pediatric intracochlear schwannomas are benign neoplasms arising within the boundaries of the inner ear, causing sudden or rapidly progressive hearing loss, dizziness, and tinnitus.
- A single report of pediatric intracochlear schwannomas was identified in the published literature.
- In this report, two additional cases of intracochlear schwannomas in pediatric patients with no signs of neurofibromatosis type II are described. Medical history and symptom progression have been reviewed together with imaging findings and management.
- Correct diagnosis should be achieved through an MRI scan with gadolinium contrast and confirmed after at least 3 months.
- The choice of management should rely primarily on the long life expectancy and growth rate of the lesion, as surgical removal of intracochlear schwannomas requires partial or total cochlear demolition, resulting in vestibular dysfunction and cochlear implant positioning preclusion. Close clinical and radiological follow-up with serial MRI scans allows to evaluate symptom progression and growth rate.

Patient 1

An 11-year-old male child was admitted complaining of progressive right-sided hearing loss that had worsened over the previous 6 months in the absence of tinnitus, dizziness, and facial weakness.

Otoscopy was unremarkable. The left ear pure tone audiometric test showed normal results, whereas a ski-sloped hearing loss was found in the right ear, with a pure tone average (PTA) of 55 dB; word recognition testing reached 70% discrimination at 60-70 dB, and a roll-over phenomenon was present at 80 dB. No identifiable waves were shown by auditory brainstem response (ABR), and otoacoustic emissions were absent in the right ear. Vestibular evaluation was normal.

CT scan was unremarkable. MRI with gadolinium revealed focal enhancement in the basal turn of the right cochlea in T1-weighted sequences with a corresponding signal loss on T2 images; findings were confirmed at a 6-month follow-up MRI and a definite diagnosis of ICS was made (Fig. 1). Subsequent MRI was performed after 12 months with stable findings. The patient did not have a family history of NF2, and a genetic consultation did not find any sign of NF2. The parents and the patient agreed upon a conservative approach consisting of a "wait and scan" strategy.

Patient 2

A 10-year-old male patient was admitted, complaining of sudden left-sided hearing loss in absence of tinnitus, dizziness, and facial weakness. The child had a clinical history of surgical excision of thoracic extra-pulmonary neuroblastoma. Otoscopy showed bilaterally normal tympanic membranes. Pure tone audiometry revealed left-sided anacusis and normal PTA in the right ear. The patient did not benefit from infusive or intratympanic corticosteroid therapy. Vestibular evaluation was unremarkable.

CT scan revealed normal anatomy of ossicles and labyrinth. MRI demonstrated focal enhancement in the middle and apical turns of the left cochlea in T1 sequences with a corresponding signal void in T2 sequences. The patient did not have a family history of NF2, and a genetic consultation excluded NF2. A putative diagnosis of ICS was confirmed after 3 months, as a second MRI showed unvaried findings (Fig. 2). The patient and his parents agreed upon a "wait and scan" strategy. Subsequent MRIs performed after 12 and 24 months showed stable findings.

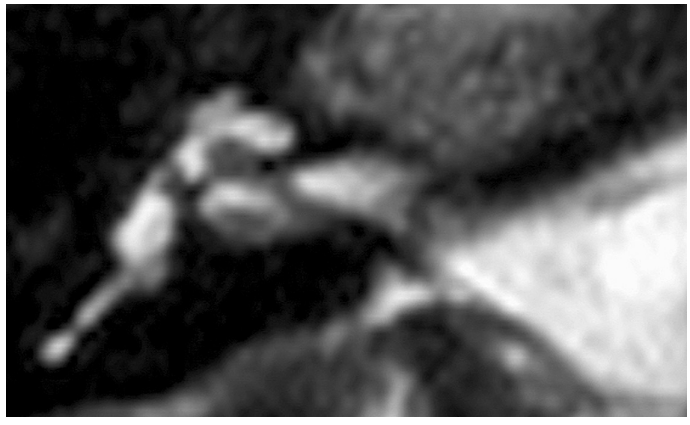

Figure 1. Patient 1, Axial MRI images of the right inner ear and internal acoustic meatus. T2 sequences show a signal void in the basal perimodiolar cochlear region.

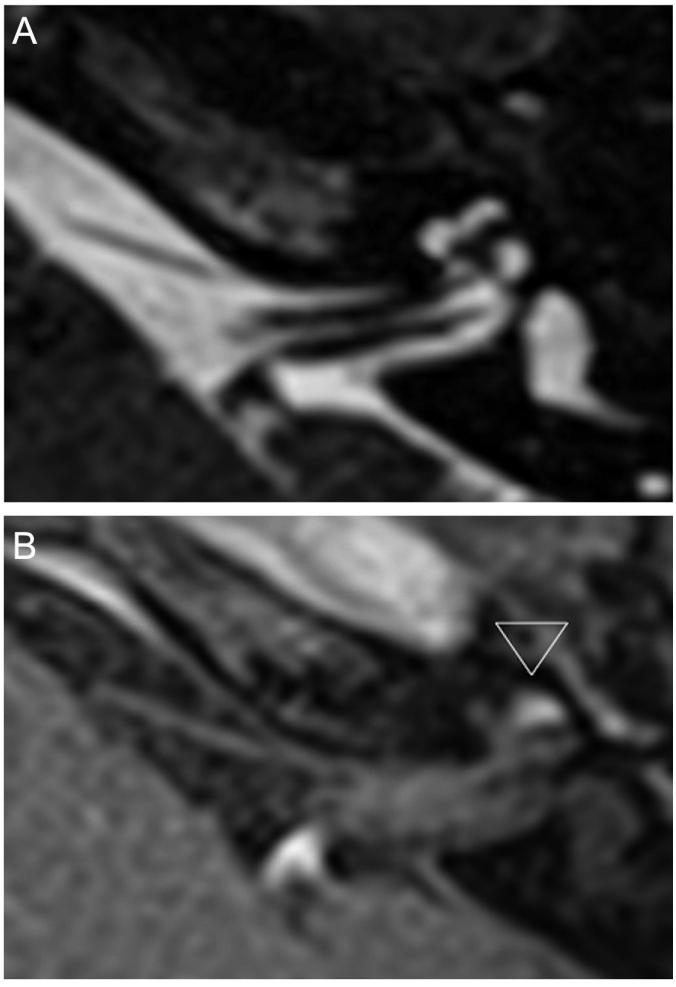

Figure 2. (a-b) Patient 2, Axial MRI images of the left inner ear and internal acoustic meatus. T2 sequences show a signal void in the middle/apical cochlear region (a). T1 sequences after gadolinium administration show an area of signal hyperintensity (arrowhead) in the middle/apical cochlear region (b).

Literature Review Results

Over the period January 2000-June 2024, three pediatric ILSs were reported in the English literature. In 2013, Van Abel et al described the case of a 15-year-old female patient, affected by Neurofibromatosis type 2, with evidence of a unilateral intravestibular lesion. After initial follow-up, she underwent gamma knife stereotaxic radiosurgery.[7] Lee et al, in 2019 mentioned the case of a 15-year-old patient, but no other clinical and radiological information were reported.[8] A single case of pediatric ICS was described by Neault et al[6]

Data Analysis

Data analysis resumes the pooled data of the latter case reported in literature[6] and authors' patients (Table 1). Overall, 3 cases of pediatric ICSs with no sign of NFII were considered: 2 patients were male and one was female. The mean age at diagnosis was 11.3 years old. The most common symptom at presentation was progressive sensorineural hearing loss (66%); sudden hearing loss was present in one case (33%). The mean duration of symptoms duration before diagnosis was 7 months (min 6 – max 12). The mean pure tone average at diagnosis was 74.2 dB. Intracochlear location was in the basal turn in 2 cases and in the apical and middle turns in the third patient. All cases initially underwent a "wait and scan" strategy. The mean follow-up time was 23.3 months.

DISCUSSION

Intralabyrinthine schwannomas originate from the perineural Schwann cells of the vestibulocochlear nerve in the osseous labyrinth without outer extension.[9,10] Presenting symptoms usually include unilateral progressive SNHL, tinnitus, vertigo, imbalance, and aural fullness; poor speech recognition ability is also very common.[1,3] Intracochlear schwannomas (ICSs) are a subtype of ILS completely located in the lumen of the cochlea arising from the modiolar region. Kennedy et al in 2004 proposed the first ILS classification.[1]

Over the last 20 years, only three pediatric ILSs were reported in the literature, including a single case of ICS described by Neault et al. They reported a 13-year-old female patient with sporadic ICS. The patient presented with rapidly progressive hearing loss and, after an initial follow-up, underwent surgery resulting in complete removal of the lesion.[6]

In this report, two additional cases of ICS in pediatric patients are described. The following results summarize the analysis of pooled data, gathered from Neault et al[6] and Authors' patients (Table 1): mean age was 11.3 years old (range 10-13 years), including two males and one female. Two patients displayed left-side involvement, whereas in one case the right side was involved. The most common symptom at presentation was progressive SNHL (66%), and sudden hearing loss was present in one case. The mean PTA at diagnosis was 74.2 dB. The range of symptoms duration at the time of diagnosis ranged from 2 to 12 months. Neither dizziness nor vertigo were reported, with bilateral normal vestibular function in the authors' patients—no information was provided by Neault et al.[6] The lesion was located in the basal turn in two cases and in the apical and middle turns in the third patient. All cases initially underwent a "wait and scan" strategy. The mean follow-up time was 23.3 months. One case displayed an

Table 1. Pooled Data Gathered from Neault et al[6] and Authors' Patients Affected by Intracochlear Schwannomas (ICS)

	Age at Diagnosis (years old)	Sex	Side	Symptoms	Symptoms Duration before Diagnosis	Location within Cochlea at Diagnosis	Hearing (PTA) at Diagnosis	Follow-up	Management
Patient 1	11	M	Right	Progressive HL	6 months	Basal turn	55 dB (stable)	12 months (stable)	Wait and scan
Patient 2	10	M	Left	Sudden HL	2 months	Middle and apical turns	120 dB (stable)	24 months (slight growth)	Wait and scan
Patient 3 (Neault et al[6])	13	F	Left	Progressive HL	12 months	Basal turn	47.5 dB (worsening)	34 months (growth)	Wait and scan + Surgery
Mean	11,3	-	-	-	7 months	-	74.2 dB	23,3 months	-

HL: hearing loss; PTA: pure tone average.

Table 2. MRI Findings of Intracochlear Schwannoma (ICS) and Differential Diagnosis [4,11,13]

Type of Lesion		T1-Weighted	T1-Weighted Post-Gadolinium	T2-Weighted Images	Follow-up
Intracochlear Schwannoma		Isointense	High contrast enhancement	Sharply delineated signal intensity loss in the high-intensity intralabyrinthine fluid	Unvaried
Acute labyrinthitis		Isointense	Enhancement most commonly affects the complete cochlea or the vestibular system	Unvaried	Enhancement becomes weaker or may disappear
Labyrinthitis ossificans	Fibrous stage	Isointense	Enhancement	Usually unsharp signal intensity loss	Possible progression to ossification
	Ossific stage	Isointense	No enhancement		Unvaried
Hemorrhage		High signal intensity	No enhancement	Variable	Hypointensity
Lipoma		High signal intensity	No enhancement	Hypointensity on fat-suppression sequences	Unvaried

evident expansion of the intracochlear lesion on MRI follow-up, leading to surgical removal of the ICS (Neault et al.).[6]

MRI with gadolinium contrast allows the detection of small intracochlear lesions (<2 mm) and an accurate description of their location, shape, signal intensity, and relation with adjacent structures and therefore represents the gold standard for the diagnosis of ILS.[1,4] Some lesions can initially mimic ICS both clinically and on contrast-enhanced MR imaging. These include acute labyrinthitis and labyrinthine hemorrhage, which should be considered in the differential diagnosis. Time-related signal modification in the case of intracochlear/intralabyrinthine lesions should be evaluated with a new exam after at least 3 months, and signal findings in ILS/ICS are meant to remain stable. Table 2 depicts the main MRI features of intracochlear lesions.[4,11-13] Radiology should always be paralleled by clinical and audiological findings before establishing a definite diagnosis.[14]

Therapeutic options for ILS/ICS include: "wait and scan" strategy, surgery, and radiotherapy [1,2,5] but unanimous consent on therapeutic guidelines in pediatric age is lacking. Surgical removal is generally considered worthwhile in the case of evidence of tumor growth into the internal auricular canal (IAC) or into the middle ear, intractable vestibular symptoms, or concern about diagnosis.[1,2,14,15] In children, it is mandatory to evaluate acoustic and vestibular functions bilaterally, as the best therapeutic option and its timing should be chosen considering the possibility of future contralateral hearing loss or vestibular dysfunction.

It is essential to consider how a different location of the lesion within the labyrinth affects the chance of rehabilitation with cochlear implants (CIs). Total surgical removal of a pure intracochlear schwannoma can be performed only through a subtotal or total cochlear demolition. Along with both clinical and audiologic indications, a cochlear implant can also be attempted without tumor removal. Laborai et al in 2022 described the cases of two adult patients: the first patient was affected by an intravestibulocochlear schwannoma, whereas the second one was affected by ICS; both underwent CI positioning without tumor removal in order to preserve the cochlear anatomy.[15] In a pediatric case, a prolonged follow-up with MRI should be performed, with obvious concerns regarding possible artifacts due to the array and consequent difficulties in monitoring tumor growth.

In the case of purely intracochlear lesion, vestibular function should be evaluated and is expected normal, as in the two patients reported. The vestibular dysfunction that invariably follows tumor removal should also be considered in therapeutic planning, particularly in pediatric cases considering their life expectancy.

It is the authors' opinion that surgical removal of an ICS in a child with a long life expectancy should be balanced against the possibility of tumor growth and contralateral hearing and vestibular status. For the same reasons, positioning of a CI without tumor removal in this context is not advisable.

Therefore, pediatric patients with ICS without invalidating vestibular symptoms should undergo a "wait and scan" strategy with serial MRI[1,2,5] and the decision on surgical removal should be deferred until evidence of tumor growth.

In the authors' opinion, there is no role for radiation therapy in pediatric patients with intralabyrinthine schwannomas.

CONCLUSION

In conclusion, ICS is extremely rare in the pediatric population. Correct diagnosis should be achieved through an MRI scan with gadolinium contrast and confirmed after at least 3 months. Consent on pediatric therapeutic guidelines is lacking. The choice of management should rely primarily on the long life expectancy and growth rate of the lesion, as surgical removal of ICS requires partial or total cochlear demolition, resulting in vestibular dysfunction and CI positioning preclusion. Close clinical and radiological follow-up with serial MRI scans allows to evaluate both symptom progression and rate of growth, in order to provide the patient with the best therapeutic option.

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Informed Consent: Informed consent was obtained from all patients whoagreed to take part in the study.

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