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SINGLE SIDED DEAFNESS & ASYMMETRIC HEARING LOSS

Case of medulloblastoma in child presented with unilateral sudden sensorineural hearing loss

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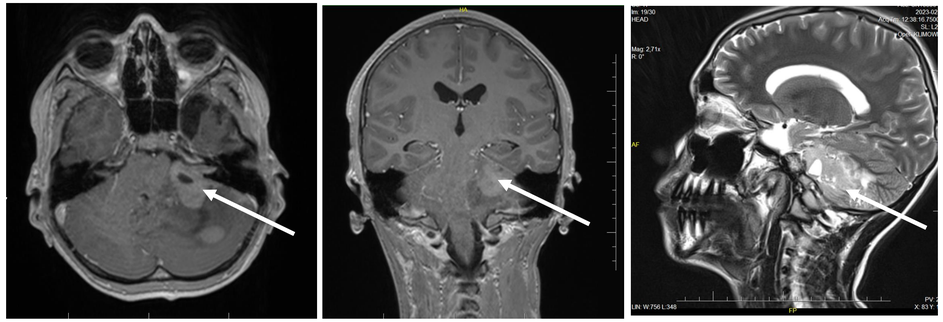
Abstract

Medulloblastoma accounts for 1.1% of all primary central nervous system tumors in all ages and up to 11.9% of those in adolescents under the age of 15 years. It originates in the cerebellum, in the posterior fossa. Anaplastic medulloblastoma is a variant characterized by aggressive behavior. The standard therapeutic management of medulloblastoma is surgery accompanied by cranio-spinal radiotherapy and multi-modal chemotherapy. Neurological signs of increased intracranial pressure and ataxia are known presentations of posterior fossa tumors. Medulloblastomas typically are associated with headaches, nausea, balance issues. Sudden sensorineural hearing loss (SSHL) as a sole symptom caused by medulloblastoma is extremely rare.

MRI with contrast (February 2023):
Extensive nodular lesion - dimensions approx. 57x42x28mm, located in the left cerebellar hemisphere, encompassing the left cerebellopontine angle, entering the canal of the internal auditory nerve, causing a reduction of the fluid space around the VII/VIII nerve complex and a reduction of the cerebellopontine angle cistern and tightening of the pericerebral fluid space at the level of the left cerebellar hemisphere



Aims

We present a rare case of medulloblastoma in 13-year-old boy with unilateral left-sided SSHL with tinnitus. Patient did not exhibit any additional neurological manifestations during admission and did not report any other complaints.
His illness may had begun two years previously when the patient was consulted neurologically due to weakness of the lower limbs, falls and periodic headaches after exertion (the symptoms disappeared spontaneously in 2021 and were diagnosed as psychosomatic).

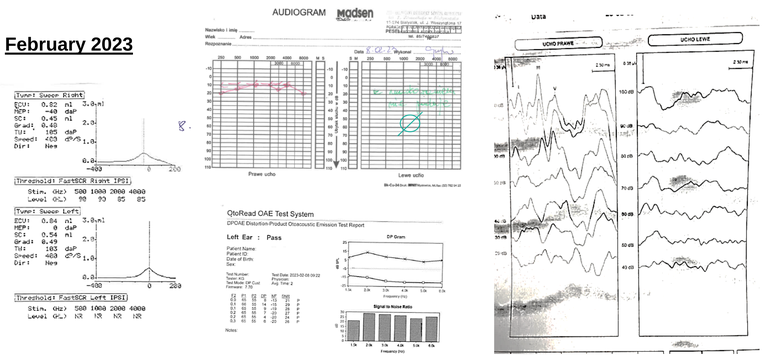
Methods and Materials

Audiological Evaluation: the boy underwent a comprehensive audiological evaluation: pure-tone audiometry, DPOAE otoacoustic emissions, Impedance audiometry, Auditory Brainstem Response (ABR) test
ENT Consultation: possible middle ear pathology was excluded
Consideration of Viral/Bacterial Infections: viral/bacterial infections were excluded
A neurological and ophtalomologic consultation: without neurological symptoms, bilateral papilledema
Imaging studies (Contrast-enhanced MRI): the poster fossa tumor was diagnosed

Results

Surgery: The primary treatment was surgical resection (complete resection was not feasible)
Histopathological Examination: Diagnosis was confirmed through surgical resection, followed by microscopic examination: Anaplastic medulloblastoma - Sonic Hedgehog (SHH) subtype p53 wild type
Radiation Therapy: Post-surgical radiation was used to target remaining tumor cells and prevent recurrence
Chemotherapy: Chemotherapy was used to September 2024
Prognosis: The progression of the remaining tumor and its spread to critical areas like the spinal cord (tumor spread to the pia mater) are associated with a poor overall prognosis

February 2023



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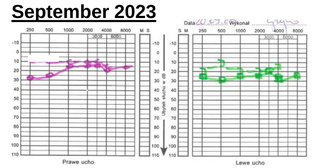

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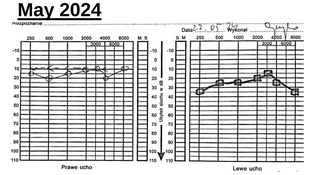

Conclusion

Recognizing the cause of SSHL and determining whether it is cochlear or retrocochlear in origin is crucial for guiding appropriate management and treatment.
Intrinsic brain tumor should be considered as a potential cause of unilateral sudden sensorineural hearing loss, as it may be easily missed leading to a delay in appropriate treatment.
Urgent radiological diagnostic is crucial for identifying and managing conditions that can lead to life-threatening neurological sequelae.
Treatment of CNS tumor may lead to improvement in hearing if the auditory nerve is preserved.

References

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