A PEDIATRIC MEDULLOBLASTOMA PRESENTING AS AN ISOLATED UNILATERAL SENSORY NEURAL HEARING LOSS.

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ABSTRACT

Medulloblastoma is one of the commonest brain tumours in the paediatric population, where they commonly present with increased intracranial pressure related symptoms. We report a child with medulloblastoma who presented with isolated right-sided unilateral sensorineural hearing loss (SNHL) for 4 months prior to diagnosis. He was found to have a large cerebellopontine angle tumour extending to foramen of Luschka and internal auditory canal. Microscopic pathology findings were consistent with medulloblastoma. Based on MEDLINE search and to the best of our knowledge, this paper is the first in the literature of pediatric medulloblastoma presented with isolated unilateral SNHL.

INTRODUCTION

The differential diagnosis of pediatric unilateral sensorineural hearing loss (SNHL) mostly focuses on syndromes and inner ear abnormalities. Unlike vestibular schwannomas, pediatric posterior fossa tumors rarely present with isolated SNHL. The question of obtaining dedicated brain imaging in this group of patients remains controversial [1]. Our pediatric medulloblastoma patient exhibited isolated SNHL as the only symptom for few months prior to progression of his clinical picture.

CASE REPORT

A healthy 7 year old male child presented to pediatric otolaryngology (ORL) clinic with right–sided hearing difficulty of 3-4 months duration. No other neuro-vestibular symptoms were present. On examination, the seventh cranial nerves' function was normal bilaterally,. An audiogram revealed a profound SNHL on the right side (figure 1). Computed tomography (CT) of the petrous temporal bone was arranged. It reported no abnormalities on either side (figure 2). Shortly after the completion of the CT temporal bones, the patient travelled out of the country with the family for a few weeks; and on the way back home, patient presented to the emergency room with symptoms and signs of increased intracranial pressure. CT and magnetic resonance imaging (MRI) of brain and spine revealed obstructive hydrocephalus, a large cerebellopontine angle mass extends into the right internal auditory meatus (IAC) (Figure 2), and a 3 mm metastatic deposit at the level of thoracic 11 spinal cord.

The Patient was admitted to pediatric intensive care unit for monitoring that night. The following morning in the operating room, the procedure started with placement of an external ventricular drain in the supine position. The patient was positioned prone and sub occipital craniotomy was carried out. Intraoperative neurophysiological

monitoring included SSEP's, MMEP and Cranial Nerve 7 monitoring. Cranial nerve VIII was found infiltrated with tumor throughout its entire course. The right VII cranial nerve was also infiltrated with the tumor; and microscopic disease was left along its course. Initial Frozen Section was consistent with a medulloblastoma.

Postoperatively the patient woke up with a right sided Grade 3 House-Brackman cranial nerve VII palsy, which improved over few weeks. External ventricular drainage was weaned off over the course of one week. The patient underwent proton beam radiotherapy and chemotherapy after the final pathology was confirmed as a medulloblastoma (figure 3). He was thereafter followed up on a regular basis by paediatric oncology team. The patient's condition deteriorated over the following year due to multiple metastases, he was treated palliatively and died within 15 months of presentation.

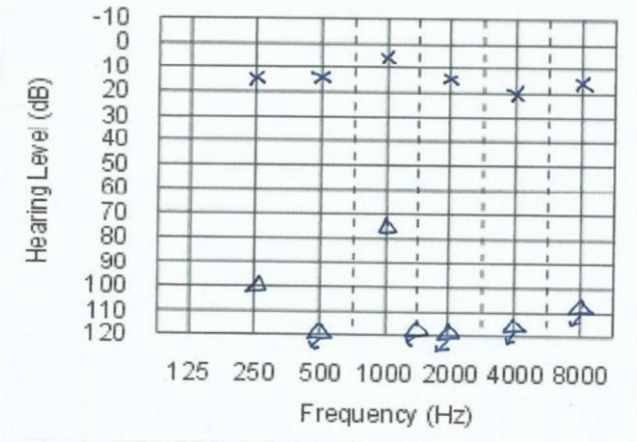

Figure 1: Pure tone audiogram demonstrates a profound sensorineural hearing loss of the right side.

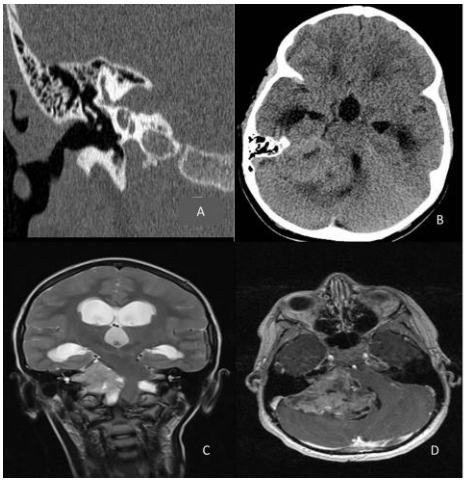

Figure 2: A) computed tomography of the right temporal bone reveals normal anatomy, B,C,D) axial CT head, coronal T2 weighted sequence head MRI , gadolinium enhanced T1 weighted sequence head MRI respectively, demonstrate hydrocephalus and right cerebellopontine angle tumor with extension to the right internal auditory meatus.

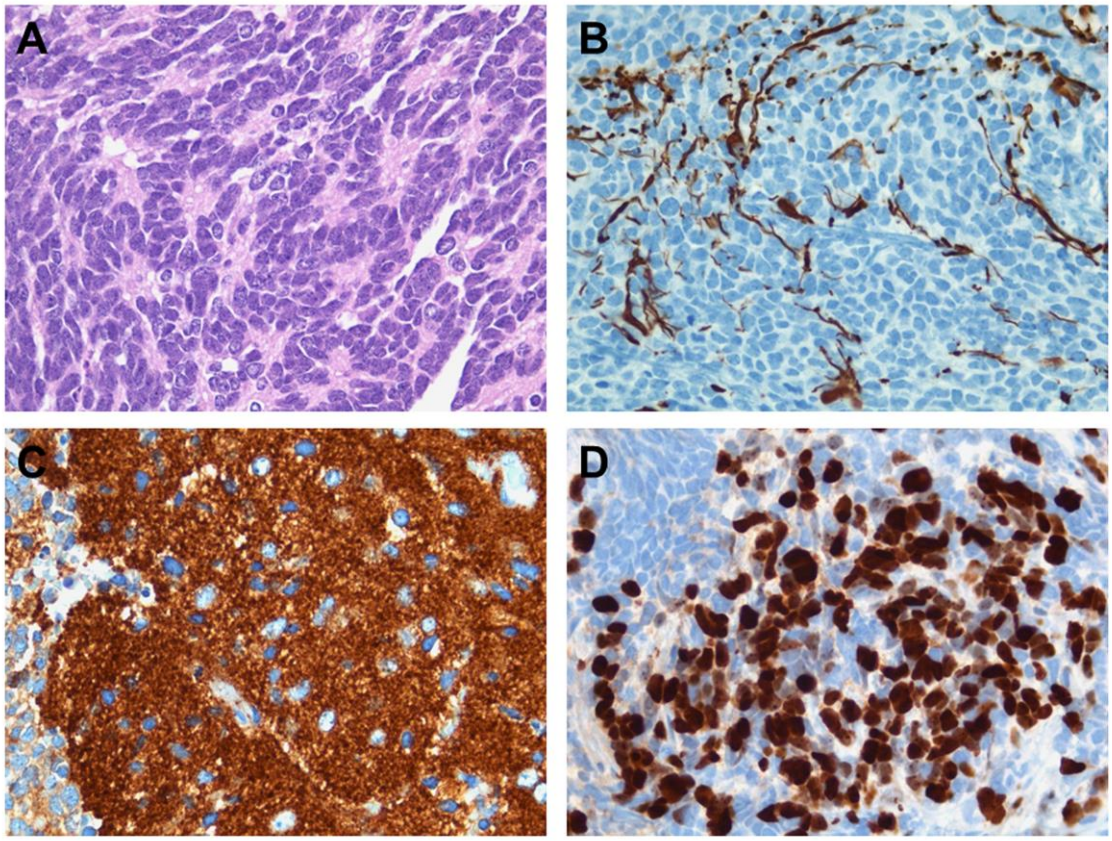

Figure 3: Photomicrographs of the neoplasm exhibiting A) a sheet of undifferentiated cells with marked hypercellularity and frequent neuroblastic (Homer Wright) rosettes, B) sparse cells immunoreactive for GFAP, C) focal immunoreactivity for Synaptophysin, and D) high Ki-67 labeling index of proliferation. Original magnification x 400 (A-D).

DISCUSSION

Pediatric SNHL is estimated to be 6 in 1000[2]. There is however no consensus on the role of imaging (MRI or CT) in unilateral SNHL in children [1]. Some studies report that up to 35% of new onset SNHL in children, have inner ear or brain abnormalities on imaging[3]. However, an ideal algorithm for imaging in children with unilateral SNHL is still lacking [2]. Both MRI and CT have their advantages and disadvantages, however, multiple recent studies are advocating a dual modality approach regardless of the order [4, 5]. In children older than 6-8 years with profound SNHL, and where anesthesia may be avoided, MRI may be considered as the first line modality[6].

With the exception of inner ear abnormalities that cause unilateral sensorineural hearing loss (SNHL) and vestibular schwannomas (VS), it is uncommon for posterior fossa tumours to present with isolated SNHL. VS particularly in neurofibromatosis 2 (NF-2) patients; represent the commonest tumour pathology of unilateral or bilateral SNHL before 15 years of age[7].

Medulloblastoma (MB) patients usually present with symptoms and signs of increased intracranial pressure such as headache, nausea, vomiting, and cerebellar dysfunction[8].MB has been reported to present with bilateral hearing loss in pediatric population[9, 10]; however, MB and other intrinsic brain tumors rarely cause

isolated SNHL. Few cases are reported describing this rare presentation. Merino Galvez et al had published the first case back in 1994; of a 25 year old patient who presented with sudden onset SNHL and was diagnosed with desmoplastic MB[11]. Later on, few more cases were published, but were all adult patients.[8, 12-14] Other intrinsic brain tumors like juvenile pilocytic astrocytoma (JPA)[15-17], rabdomyoma [18] and arachnoid cyst [7] are reported to cause SNHL in pediatric population.

Neely et al. hypothesized that the SNHL associated with VS could result from either direct VIII nerve compression, vascular involvement, biochemical changes in the inner ear, or decrease in the number of nerve fibres[11]. As the auditory neuronal pathway is bilaterally represented, SNHL in our case has to affect afferent nerve fibres distal to the cochlear nucleus[8] where this effect could be due to compression of VIII cranial nerve brainstem segment before it joins the ipsilateral cochlear nucleus or along the cisternal segment.

In conclusion, and to the best of our knowledge, our paper is the first to report unilateral SNHL as the initial sole presentation of a MB in pediatric population. This and other cases of different brain tumors presenting similarly raise the question if dedicated neuronal imaging should be included in the initial work up of SNHL in pediatric patients.

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