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Peripheral facial palsy, the only presentation of a primitive neuroectodermal tumor of the skull base

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ABSTRACT

INTRODUCTION: Peripheral facial palsy is rarely caused by primary neoplasms, which are mostly constituted of tumors of the central nervous system, head and neck, and leukemia.
PRESENTATION OF CASE: A 2-month-old male infant presented with asymmetric facial expression for 3 weeks. Physical examination revealed suspicious findings of right peripheral facial palsy. Computed tomography of the temporal bone revealed a suspicious bone tumor centered in the right petrous bone involving surrounding bones with extension into the middle ear cavity and inner ear. Subtotal resection of the tumor was performed due to crucial structures adjacent the mass. Histopathology and immunohistochemistry of the resected tumor was consistent with primitive neuroectodermal tumor.
CONCLUSION: We report a rare case of a primitive neuroectodermal tumor located at the skull base presenting with only peripheral facial palsy.

1. Introduction

The cause of peripheral facial palsy in the pediatric population is mostly idiopathic. According to previous studies, 16–50% of all peripheral facial palsy cases in children are unknown [1,2]. Identified causes of peripheral facial palsy in children include trauma, infection, congenital anomalies, and neoplasms [1]. Peripheral facial nerve palsy caused by a primary neoplasm is rare, and is mostly due to tumors of the central nervous system (CNS), head and neck, or leukemia [3]. Reports of tumors of the skull base presenting with only signs of peripheral facial palsy is scarce. We report a rare case of right facial palsy in a 2-month-old infant due to suspicious compression of the right facial nerve by a tumor of the skull base without involvement of the CNS, which was identified as primitive neuroectodermal tumor (PNET).

2. Case report

A 2-month-old male infant visited the Department of Pediatrics of Inha University Hospital due to asymmetric facial expression which had first appeared 3 weeks before. He was born at 38 weeks of gestational age and 3.08 kg of birth weight. He did not have asymmetric facial expression at birth and was healthy since birth. Family history was also unremarkable.

Initial vital signs were normal and his mental status was alert. Physical examination of the general systems was unremarkable. On neurologic examination, pupils were isocoric with prompt light reflexes. His oral angle movement was asymmetrical and nasal fold and forehead crease was shown in only the left face when he was crying. He was unable to close his right eyelid completely when he was crying, while closure of both eyelids was observed in resting conditions (Fig. 1). Conjunctival hyperemia was also observed in his right eye. Motor strengths were normal in all extremities and sensory was intact. Biceps and ankle jerk reflexes were normal and no pathologic reflexes were detected.

Brain magnetic resonance imaging (MRI) revealed a well-enhanced expansible mass of 3 cm in diameter in the right petrous bone compressing the right temporal lobe (Fig. 2). The mass involved the right cavernous sinus and the Meckel's cave destructing the clivus. The mass was shown to medially bulge out to the masticator space through the skull base extending to the neck level of the infratemporal fossa. Bulging to the posterior aspect of the internal auditory canal, suspicious nerve compression, and combined right otomastoiditis was also observed. Computed

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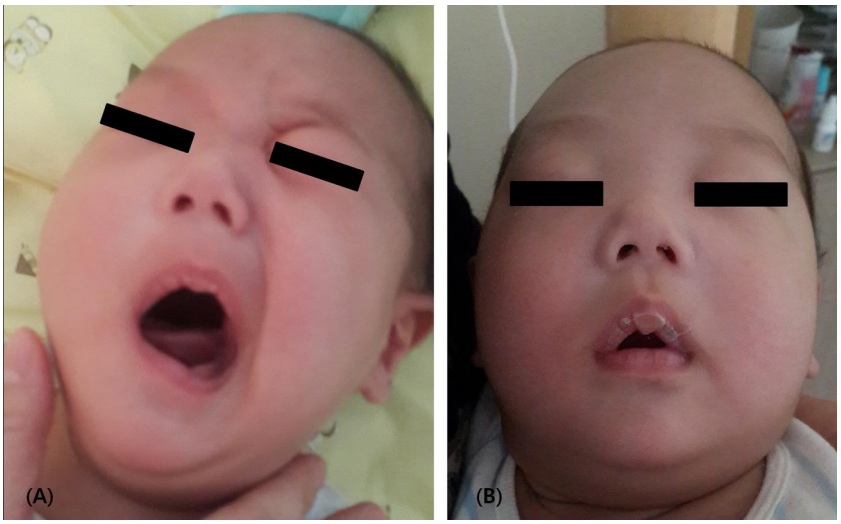

Fig. 1. Facial expression of the patient at presentation. (A) The patient's oral angle movement was asymmetrical, and the nasal fold and forehead crease was shown only in the left face during crying. (B) Symmetrical facial expression was observed during rest.

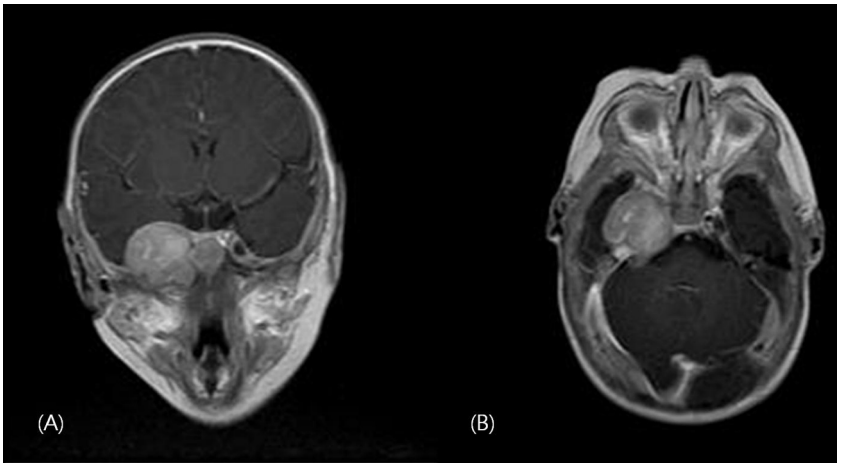

Fig. 2. Gadolinium-enhanced T1 weighted magnetic resonance images of the brain. Axial images (A) and coronal images (B) show a well enhancing outbulging mass of approximately 3 cm size in the right petrous bone, compressing the right temporal lobe.

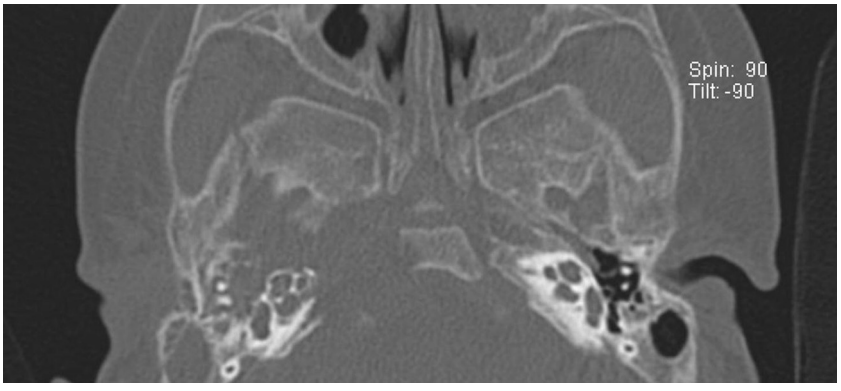

Fig. 3. Computed tomography of the temporal bone. An expansile osteolytic bone tumor located in the right central skull base, involving the right temporal bone and clivus, and extending to the middle ear cavity and inner ear structures. Suppurative right otomastoiditis with multifocal outer table erosions was also observed.

tomography of the temporal bone showed a suspicious bone tumor centered in the right petrous bone involving surrounding bones (Fig. 3). Extension of the tumor into the middle ear cavity and inner ear structure was also observed. Auditory brainstem-evoked response threshold was normal in the right ear, while the threshold of the left ear was 30 dB nHL, which was consistent with left ear deafness. Bone scan of the whole body showed no metastasis in other bone lesions.

Total resection of the tumor was attempted by the Department of Neurosurgery. However, total resection of the tumor was