CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 19 (2016) 47–50

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

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

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ARTICLE INFO

Article history:
Received 27 November 2015
Received in revised form
12 December 2015
Accepted 13 December 2015
Available online 17 December 2015

Keywords:
Facial palsy
Primitive neuroectodermal tumor (PNET)
Skull base neoplasm

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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http://dx.doi.org/10.1016/j.ijscr.2015.12.019

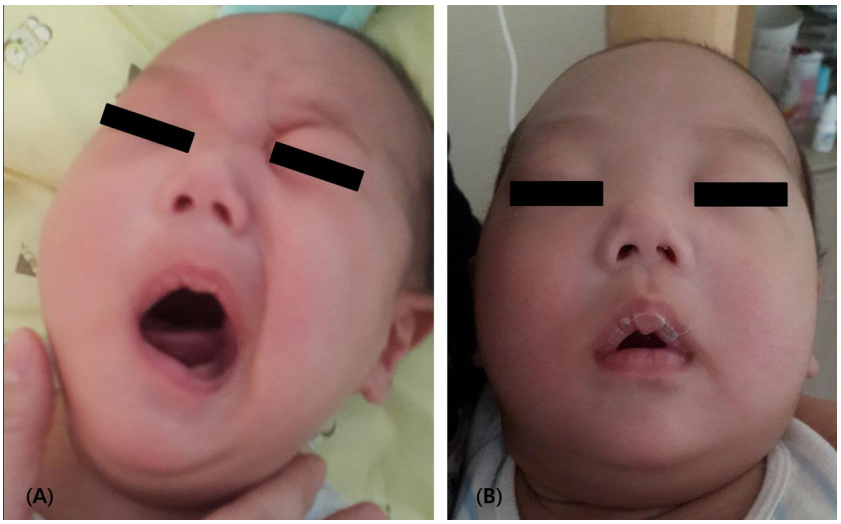

Fig. 1. Facial expression of the patient at presentation. (A) The patient's oral angle movement was asymmetrical, and the nasal fold and forehead crease was shown only in the left face during crying. (B) Symmetrical facial expression was observed during rest.

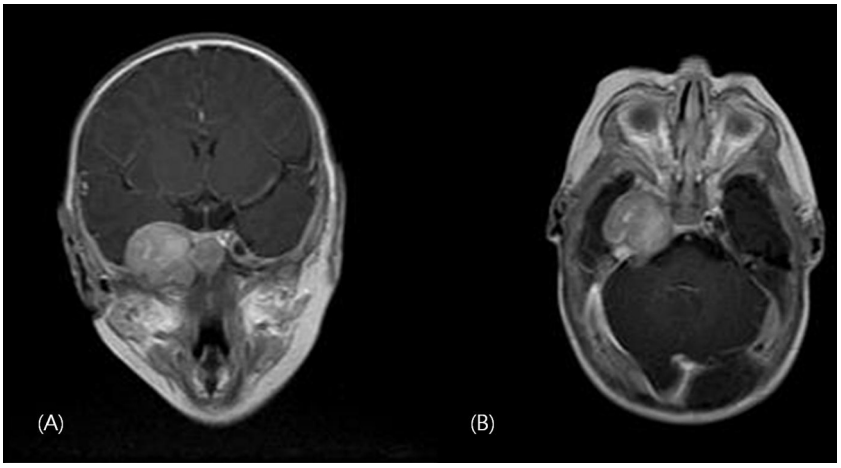

Fig. 2. Gadolinium-enhanced T1 weighted magnetic resonance images of the brain. Axial images (A) and coronal images (B) show a well enhancing outbulging mass of approximately 3 cm size in the right petrous bone, compressing the right temporal lobe.

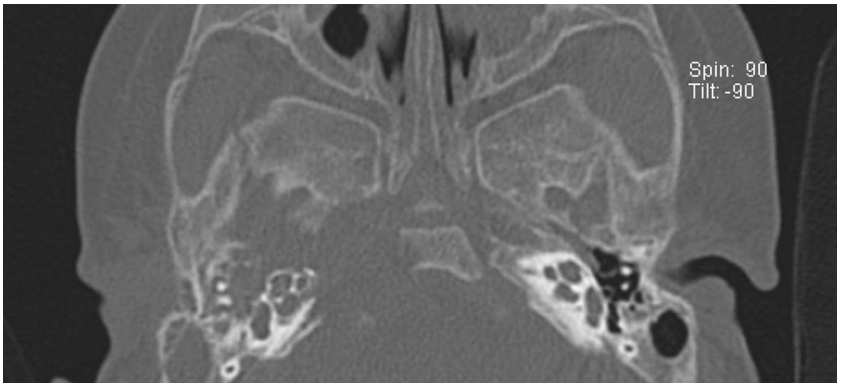

Fig. 3. Computed tomography of the temporal bone. An expansile osteolytic bone tumor located in the right central skull base, involving the right temporal bone and clivus, and extending to the middle ear cavity and inner ear structures. Suppurative right otomastoiditis with multifocal outer table erosions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

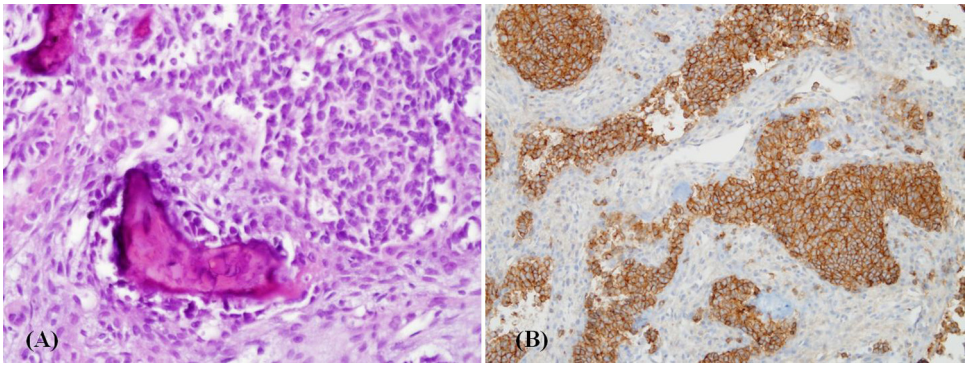

Fig. 4. Histopathologic findings of the surgically removed bone tumor of the right petrous bone (A, ×400). The medullary spaces were replaced by densely cellular fibrous connective tissue with infiltrative solid nests of malignant small round tumor cells showing abundant plasma-like cells proliferation. Immunostaining for CD99 was positive (B).

incapable due to crucial structures adjacent the mass. Histopathology of the subtotally resected tumor mainly comprised of malignant small round cell, which was consistent with sarcoma. Immunohistochemistry revealed a neural phenotype with diffuse and strong positive staining for CD99 and vimentin, which was consistent with PNET (Fig. 4).

After subtotal resection of the tumor, the asymmetrical facial expression of the patient slightly improved. The patient initiated induction chemotherapy with a combination of vincristine, cyclophosphamide, and etoposide, and is currently on continuation therapy with vincristine and cyclophosphamide (COG- AEWS0031 protocol).

3. Discussion

PNETs are malignant small cell neoplasms which develop from undifferentiated neuroepithelial cells. They predominantly occur early in life, but can develop at any age. PNETs usually occur in the cerebrum but also develop in the brain stem, spinal cord or peripheral nerves [4]. In past, the term 'PNET' was used for tumors located in the CNS (CNS-PNET). More recently, however, the PNET concept has expanded to include similar tumors located in bones and soft tissue of the body, which is referred to as peripheral primitive neuroectodermal tumor (peripheral-PNET) [5].

The diagnosis of PNET depends on histology and immunohistochemistry findings. Histological evidence of Homer-Wright or Flexner–Wintersteiner rosettes, and immunoreactivity to two or more neural markers are required for diagnosis. Ultra-structural transmission electron micrographic evidence of neural differentiation and neurosecretory granules is also helpful for the diagnosis of PNET. The CD99 and the vimentin positive staining in tumor cell is important in the differentiation of PNET from other small round cell tumors [6].

Clinical features of PNET depend on primary tumor site and extent of the disease. Headache, vomiting, seizure, eye signs, hemiparesis and hydrocephalus has been reported in patients with PNET in the cerebrum, with a mean duration of 3 months from symptom onset to diagnosis [7]. Peripheral PNET can occur in nerves, soft tissues, and bones. In one retrospective analysis, more than half of malignant peripheral PNET occurred as thoraco-pulmonary skeletal lesion, which is also known as Askin's tumor [8]. Other cases of in the analysis, malignant peripheral PNET were localized in the abdomen, pelvis, extremities, head and neck. Most of those tumors involved both bones and soft tissues. The tumor in our case was located in the skull base, without involving the CNS. Therefore, cerebral symptoms and signs were not observed in our case. We assume that the facial palsy in our case was due to the compression effect of the tumor, along with the suspicious compression of the auditory nerve shown on brain MRI causing deafness. The skull base is an uncommon location for peripheral PNET to occur primarily in infant, and there are scarce relevant reports [4].

Facial palsy is an uncommon manifestation in patients with tumors located in the skull base. According to one study that analyzed the etiology of facial palsy, only 1 out of 349 pediatric patients revealed a solid tumor, which was identified as a malignant lymphoma [9]. In a study of skull base tumors, only 4% of pediatric and adolescent patient reported complaints of facial weakness. Histopathology results of the tumors in this study were benign juvenile nasopharyngeal angiofibroma, benign nerve sheath tumor, and malignant embryonal rhabdomyosarcoma [10]. To our knowledge, there is no report of a skull base tumor of peripheral PNET presenting with only facial palsy. To date, we are the first to report a case of facial palsy due to suspicious compression of the facial nerve by a tumor of the skull base without involvement of the CNS, which was identified as PNET.

The prognosis of peripheral PNET is generally poor. About 56% of patients with localized PNET remained disease-free for 3 years from diagnosis [8]. According to one report, among 6 patients who received subtotal resection due to PNET in the head and neck region, only 50% (3/6) showed complete remission [11]. Localized PNET in the extramedullar location was less life-threatening and less aggressive and had a more chance of total resection of the tumor than PNET in the CNS, while a patient with bony infiltration and bone marrow metastasis had a poor prognosis [12]. In our case, no evidence of metastasis in other bone lesions was a good prognostic factor, while failure of total resection of the tumor was a poor prognostic factor for patient survival.

We report a rare case of facial palsy in a 2-month-old infant due to PNET located in the skull base. We are the first to report a case of malignant peripheral PNET of the skull base that presented only with facial palsy, by compression of the facial nerve by the PNET. It should be kept in mind that compression of the facial nerve by tumors of the skull base may present only with facial palsy, especially in young infants. Moreover, although peripheral facial palsies are mostly idiopathic, thorough evaluation should be performed to clarify the underlying cause as in our case.

Conflict of interest

No conflicts of interest was declared by the authors.

Funding

The authors declare that this study has received no financial support.

Ethical approval

This report was not a research study, but a case report. No ethical approval was obtained for this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report.

Author contribution

Hyung Jin Kim: drafted the manuscript.
Ben Kang: revision and proofreading of the manuscript.
Eun Young Joo: contributed in data collection.
Eun Young Kim: performed the surgery in this case and revised the manuscript.
Young Se Kwon: contributed in deriving this case, reviewing the literature, and revision of the manuscript.

Guarantor

Young Se Kwon.

Acknowledgment

This study was supported by Inha University Research Grant.

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