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What is This?
Metastatic Neuroblastoma Presenting as Recurrent Facial Paralysis

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Keywords
facial paralysis, recurrent facial paralysis, pediatric facial paralysis, neuroblastoma, metastatic neuroblastoma, temporal bone mass, rare cause of temporal bone mass, pediatric malignancy, neuroblastoma of the head and neck

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Metastatic neuroblastoma is an extremely rare cause of a temporal bone mass. We describe a 4-year-old who presented with recurrent right facial paralysis. Radiographic study and biopsies of the middle ear, bone marrow, and an abdominal mass revealed advanced stage IV metastatic neuroblastoma. To this date, there are no known reports of stage IV metastatic neuroblastoma presenting as recurrent facial paralysis.

Case Report
A 4-year-old boy presented to his pediatrician in January 2010 with right-sided facial paralysis. The patient was treated with a 1-week course of oral corticosteroids and the facial paralysis resolved. Three months later, the patient developed right-sided hearing loss, otalgia, and an abdominal mass revealed advanced stage IV metastatic neuroblastoma. To this date, there are no known reports of stage IV metastatic neuroblastoma presenting as recurrent facial paralysis.

On physical examination, the patient’s right face showed flattening of the nasolabial fold, mouth droop, and peripheral facial nerve weakness without widening of the palpebral fissure. Examination of the right ear showed a normal external auditory canal with a reddish, dull, nonpulsatile, intact tympanic membrane without surrounding bone erosion. Audiological evaluation of the right ear indicated a severely elevated pure-tone threshold and a speech recognition threshold of 100 dB and 0% speech discrimination. Audiological evaluation of the left ear was normal.

Computed tomography (CT) scan of the brain revealed a large mass within the right temporal bone with extensive intracranial invasion (Figure 1a), including the internal auditory canal with encasement of cranial nerves VII and VIII. Magnetic resonance imaging (MRI) of the brain and skull base showed infiltration of the subcutaneous tissues of the right side of the face surrounding the pinna and occlusion of the right eustachian tube proximal to the right middle ear. A middle ear biopsy was performed, revealing a soft, nonvascular, necrotic mass. Pathological examination revealed a poorly differentiated primitive neural tumor with a pink, fibrillar neuropil centered within rosette-like structures, consistent with neuroblastoma (Figure 2).

Computed tomography of the chest, abdomen, and pelvis was ordered to determine the source of the primary tumor.

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A 3.8 × 4.2 × 4.7-cm well-circumscribed mass was found anterior to the left kidney. Metastatic sites were noted in the T8 vertebral body and the right femoral neck. Biopsy of the retroperitoneal mass confirmed the presence of poorly differentiated neuroblastoma.

The patient was diagnosed with advanced stage IV neuroblastoma and underwent chemotherapy and immunotherapy. The patient received radiation therapy in the left suprarenal fossa and to metastatic sites in the T8 vertebral body and right femoral neck. Nearly 24 months after initial presentation, persistent skull-based disease was noted on MRI, and the patient’s right-sided facial paralysis remains.

Discussion
Pediatric facial nerve palsy may be congenital or acquired. Acquired causes include cholesteatomas, parotid lesions, paragangliomas, sarcomas, teratomas, vascular abnormalities, and metastatic disease. For patients with an otoscopically demonstrable mass, history of chronic otitis media, acute mastoiditis, or trauma, high-resolution temporal bone CT is recommended for evaluation of the intratemporal portion of cranial nerve VII. Magnetic resonance imaging is especially helpful in identifying brainstem pathology. Biopsy of a suspected mass can be done within the middle ear.

Neuroblastoma is the third most common pediatric malignancy, accounting for approximately 10% of all childhood cancers. Neuroblastoma arises from neural crest tissue of the sympathetic ganglia or adrenal medulla and is characterized by diverse clinical behavior ranging from spontaneous remission to rapid metastatic progression and death. Approximately 70% of patients with neuroblastoma have metastatic disease at diagnosis. Common locations include the skull, periorbital bones, bone marrow, and liver. Metastatic neuroblastoma presenting as recurrent facial paralysis, however, is rare.

The prognosis for patients with neuroblastoma is related to age at diagnosis, stage of disease, site of the primary tumor, tumor histology, and, in patients older than 1 year, regional lymph node involvement.

The International Neuroblastoma Staging System defines stage IV neuroblastoma as any primary tumor with dissemination to distant lymph nodes, bone, bone marrow, liver, skin, and/or other organs. Standard therapy for patients with stage IV disease involves induction, local control, consolidation, and treatment with biologic agents. About 80% of patients achieve complete or partial remission, although in advanced disease, remission is seldom durable.

Case reports without intervention do not require review or approval from the Memorial Healthcare System Institutional Review Board.

Author Contributions
Ariel B. Grobman, literature review, research, authorship, citation building; Lawrence R. Grobman, patient care, authorship, literature review, editing; Anne Schaefer, patient care, editing.

Disclosures
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