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Hemangioma of the Internal Auditory Canal and PHACES Syndrome: A Rare Finding in a Rare Syndrome

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PHACES syndrome represents a rare congenital anomaly with a broad spectrum of clinical manifestations. The etiology remains unclear but it occurs in approximately 2.0% of patients with facial hemangiomas.[1] PHACES syndrome is characterized by the presence of segmental facial hemangioma of infancy (H) accompanied by at least one of the following extracutaneous manifestations: posterior fossa malformation (P), arteriovenous malformation (A), cardiovascular anomaly (C), eye abnormality (E), or ventral defect including sterna clefting or supraumbilical raphe (S).[2]

The most common otolaryngological manifestations seen in the children with PHACES syndrome are hemangiomatous involvement of tympanic membrane, narrowed nares with functional nasal obstruction, hemangiomatous involvement of oral or pharyngeal mucosa, and airway hemangioma.[2]

We present a case of PHACES syndrome that has been seen in our department. Large cervicofacial hemangioma, posterior fossa malformations, and hemangioma in the internal auditory canal (IAC) were found in this patient.

In 2012, a 1-year-old girl presented with large cervicofacial hemangioma since birth with no symptoms or signs of stridor, dysphagia, nasal obstruction, or hearing loss. There was no significant prenatal, birth, or postnatal history or any family history of hearing loss. On physical examination, the external auditory canals and tympanic membranes were normal. No hemangioma was found by laryngoscopy. A computed tomography scan of the neck was performed, showing a cervicofacial hemangioma (30 × 35 × 50 mm) on the right (Figure 1A). Magnetic resonance imaging and magnetic resonance angiography were obtained (Figure 1B) to identify associated intracranial anomalies, which included a posterior fossa arachnoid cyst, a dysmorphic cerebellum, and an enhancing mass (5.4 mm largest diameter) within the right IAC consistent with hemangioma (Figure 2A and B). Although our patient had passed her newborn hearing screen, the patient was screened by otoacoustic emissions, and due to extension of hemangioma within the IAC, an auditory brain stem response was obtained and revealed normal and symmetric interpeak intervals and peak latencies increased in the right ear.

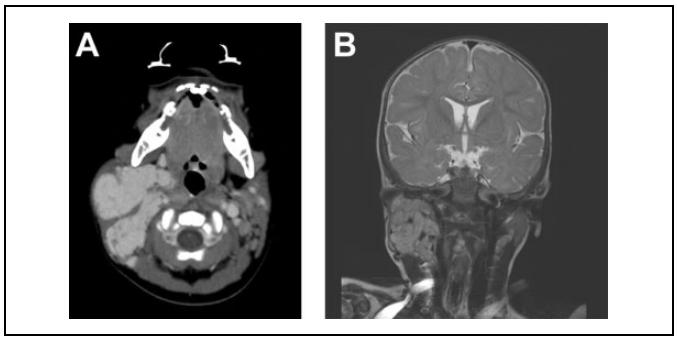

Figure 1. 2012 Axial CT scan with contrast (A) and coronal T2 MR image (B) showing a cervicofacial hemangioma on the right. CT indicates computed tomography; MR, magnetic resonance.

The infant was treated with propranolol and had significant regression of the cervicofacial hemangioma (Figure 3) with slight reduction in the hemangioma of the IAC. During the last 3 years, the patient has continued under regular follow-up and is thriving well with no speech delay or others relevant symptoms.

Vascular lesions of the IAC are extremely rare,[3] but it should be considered in any infant with a large, segmental, facial hemangioma. Observation has been recommend as the primary mode of therapy.[4] Because of the rarity of PHACES syndrome, otolaryngologists are often faced with a diagnostic challenge. It is important to understand its fundamental elements and how they affect the patients seen in our field.

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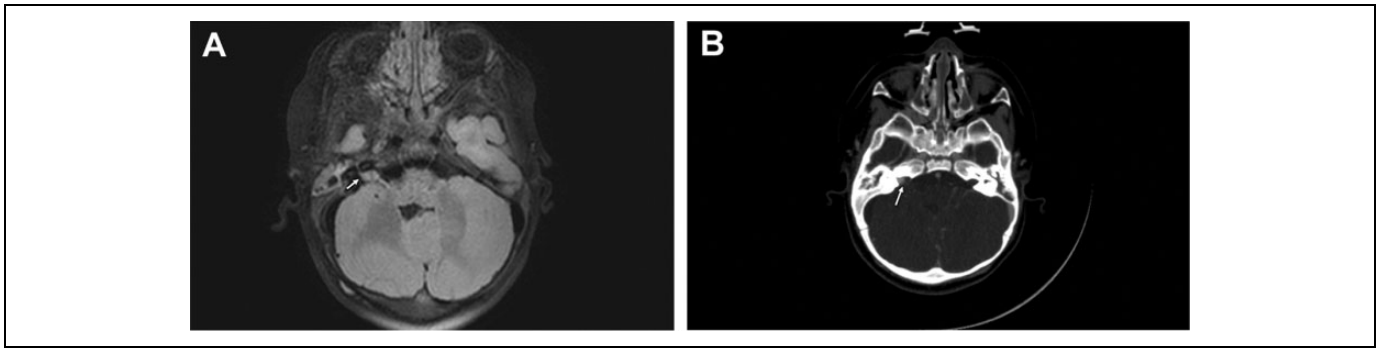

Figure 2. 2012 Axial T2 Flair MR image (A) and axial CT scan with contrast (B) showing an enhancing mass within the right internal auditory canal consistent with hemangioma. CT indicates computed tomography; MR, magnetic resonance.

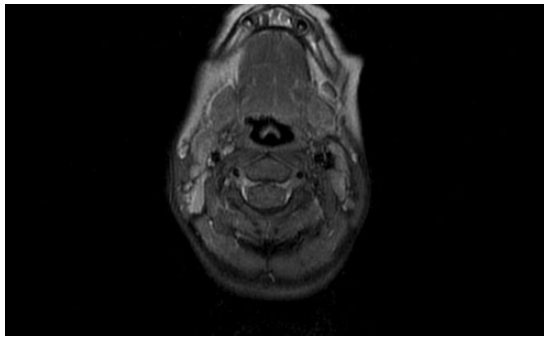

Figure 3. 2016 Axial T1 MR image with contrast showing significant regression of the cervicofacial hemangioma. MR indicates magnetic resonance.

Declaration of Conflicting Interests

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