

Progressive Facial Paralysis Caused by Heterotopic Ossification of the Stylohyoid Ligament

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A 56-year-old man with a history of a right skull base fracture from a fall in the military 30 years ago presented to clinic with 6 months of progressive right-sided facial paralysis and a 1-year history of worsening odynophagia and dysphagia. He had been previously diagnosed at an outside facility with Bell's palsy. Upon examination, the patient had House-Brackmann grade IV facial palsy on the right, tenderness to palpation over the right mastoid tip, and a palpable firm mass below the right mandible with fixation of the hyoid bone. Computed tomography of the neck showed a calcified mass extending from the right posterior skull base to the hyoid (**Figure 1**). There was narrowing of the facial canal as the nerve exits the stylomastoid foramen.

The patient underwent transcervical resection of the calcified mass and mastoidectomy with facial nerve drill-out. During the mastoidectomy, there was evidence of facial nerve impingement as it exited the skull base. Although the nerve was grossly intact, it could not be stimulated. A partial parotidectomy and level II-IV neck dissection were performed to provide access for removal of the mass (**Figure 2**).

Eighteen months postoperatively, the patient still has House-Brackmann grade IV on the right. His dysphagia and odynophagia improved but did not completely resolve.

University of Florida Institutional Review Board exemption was obtained for this case report.

Discussion

Eagle syndrome is a clinical diagnosis defined by the presence of an elongated styloid process >30 mm and/or a calcified stylohyoid ligament, with incidence of a variety of

symptoms. It is classified into 2 clinical types: classic and carotid. The classic type commonly presents with a globus sensation or with unilateral pharyngeal dull pain triggered by swallowing.¹ The glossopharyngeal nerve is the most likely cranial nerve to be affected due to its course near the tonsillar fossa.² The carotid type usually presents with movement-induced neck pain and headaches, as there are compressive forces on the internal or external carotid. This can lead to ischemic symptoms, such as syncope, headaches, stroke, visual loss, and even sudden death if affecting the internal carotid, while compression of the external carotid artery leads to ipsilateral neck and face pain.^{3,4}

There are several theories to explain how Eagle syndrome develops, including congenital elongation, idiopathic calcification of the stylohyoid ligament, and ossification at the insertion at the hyoid.¹ When an enlarged styloid is found to be the causative agent for the patient's symptoms, initial treatment options include involve reassurance to the patient, analgesics, and steroid injections. Those who fail medical treatment may be offered transoral styloidectomy with or without tonsillectomy and transcervical styloidectomy, although up to 20% report persistence of symptoms postoperatively.⁵

Our patient is the first documented case of Eagle syndrome presenting as progressive facial paralysis to be described in the medical literature. The enormity of the styloid and, thus, the etiology of the patient's symptoms were due to the extraskeletal bone formation along the stylohyoid muscle. This entity, described as heterotopic ossifications, can further be categorized into 3 types: neurogenic, traumatic, and myositis ossificans progressiva. This patient's remote history of a fall indicated that this was due to traumatic heterotopic ossification. The facial paralysis was caused by

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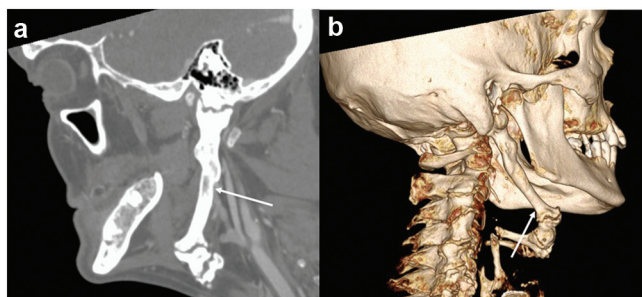


Figure 1. (a) Sagittal computed tomography scan, with the arrow showing an enlarged styloid from mastoid to hyoid bone. (b) Three-dimensional computed tomography reconstruction demonstrating a large calcified mass, denoted by the arrow, inferior and medial to the mandible on the right.

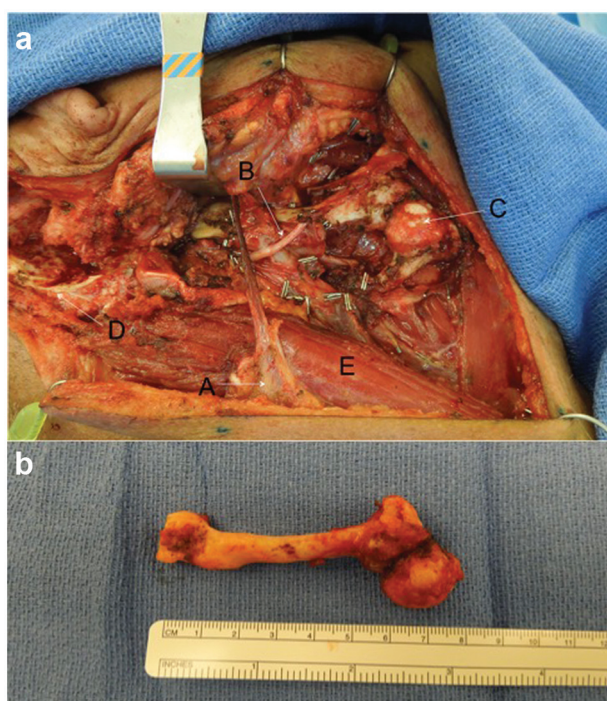


Figure 2. (a) Gross neck dissection: A, external jugular vein; B, hypoglossal nerve; C, calcified mass; D, mastoid tip; E, sternocleidomastoid. (b) Excised mass, corresponding to area labeled C.

long-standing impingement of the facial nerve at the stylo-mastoid foramen. Unfortunately, the facial nerve decompression did not improve the patient's palsy, likely due to the chronicity of its compression between the styloid and mastoid tip.

This case highlights an unusual presentation of Eagle syndrome attributed to facial palsy caused by traumatic heterotopic ossification. While this is an unlikely cause of a facial nerve disorder, it demonstrates the potential for multiple cranial neuropathies that can be associated with Eagle syndrome.

Author Contributions

Joshua P. Weiss, drafting the conception of the paper, literature analysis, writing of the case report; **Peter T. Dziegielewski**, involved in discussion of concept to draft paper, analysis of paper with comprehensive edits including final review.

Disclosures

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