

A rare case of Eagle syndrome and diffuse idiopathic skeletal hyperostosis in the cervical spine

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Published case studies involving diffuse idiopathic skeletal hyperostosis (DISH) along with symptomatic ossification of the stylohyoid ligament, or “Eagle syndrome,” are rare. In this report, we document a case of bilateral Eagle syndrome and advanced DISH in the cervical spine of an 80-year-old man who presented with severe neck stiffness and intermittent asphyxia. The key imaging and clinical features of this disorder are also described.

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KEY WORDS : Eagle syndrome, diffuse idiopathic skeletal hyperostosis, airway obstruction, chiropractic

Les études de cas publiées concernant l’hyperostose squelettique idiopathique diffuse (HSID) avec une ossification symptomatique du ligament stylohyoïdien, ou « syndrome d’Eagle », sont rares. Dans le présent rapport, nous consignons un cas de syndrome d’Eagle bilatéral et de HSID avancée du rachis cervical chez un homme de 80 ans présentant une raideur grave de la nuque et une asphyxie intermittente. Les principales caractéristiques d’imagerie et cliniques de ce trouble sont également présentées.

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MOTS CLÉS : syndrome d’Eagle, hyperostose squelettique idiopathique diffuse, obstruction des voies respiratoires, chiropratique

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Consent: The patient has provided written consent to having his personal health information, including radiographs, published.

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Figure 1. Anteroposterior open mouth radiograph of the upper cervical spine reveals thick, diffuse ossification of both stylohyoid ligaments (*).



Figure 2. Lateral cervical spine radiograph. The extensive ossification of the stylohyoid ligaments, identified by asterisks (*) extends from the skull base to the hyoid bone. A pseudoarticulation between the abnormal ossification and the hyoid bone is evident. Also observed is an 8 mm thick vertically oriented shield of ossification adjacent to the anterior vertebral bodies and disc spaces from C2 through C7 typical of DISH. The intervertebral disc spaces are typically preserved and the facet joints are normal in DISH as illustrated in this case. Of note, there is a 1 cm circular calcified mass located anterior to the C6-7 level most likely representing a benign thyroid cyst.

Case Presentation

An 80-year-old white male presented to a chiropractic clinic with a 3-year history of insidious and progressing neck stiffness and “cramping,” increased difficulty turning his head from side-to-side (e.g. while driving), and a “choking” sensation when flexing his neck. He also complained of a constant pressure in his throat as well as difficulty swallowing his saliva, to the point where he felt like he was “choking on his spit at times.” He had no problems with normal breathing, speaking, or swallowing food and he denied any symptoms suggestive of cervical radiculopathy or myelopathy. The only difficulty he reported with eating was an “inability to look down at the plate to see his food.” On examination, his cervical spine range of motion was severely limited (by 80-90%) in all directions. Flexing his neck forward beyond five degrees would elicit asphyxia. Bilateral upper limb neurologic examination, including motor, reflex, and sensory testing, was unremarkable. Cervical spine radiographs revealed complete bilateral ossification of the stylohyoid ligaments as well as advanced ossification of the anterior longitudinal ligament (flowing hyperostosis) throughout the cervical and upper thoracic spine, abutting and displacing the prevertebral soft-tissues (Figures 1-3). Based on these findings, the patient was diagnosed with Eagle syndrome and diffuse idiopathic skeletal hyperostosis (DISH).

Discussion

Symptomatic elongation of the styloid process and/or ossification of the stylohyoid ligament was first described by Eagle^{1,2} in the late 1940s and is known as *Eagle syndrome*. Elongated styloid processes or ossified stylohy-

Table 1.

Key imaging and clinical features of Eagle syndrome

Key imaging features
<ul style="list-style-type: none"> Ossification of the stylohyoid ligaments is best seen on lateral cervical and anteroposterior upper cervical radiographs
Clinical features ¹⁻⁴ : dysphagia (i.e. difficulty swallowing), sensation of a foreign body in the throat, otalgia (i.e. pain radiating to the ear), constant dull nagging ache in the throat, carotodynia (i.e. pain in the distribution of the carotid arteries), temporomandibular joint pain, glossopharyngeal neuralgia, and facial pain or headache

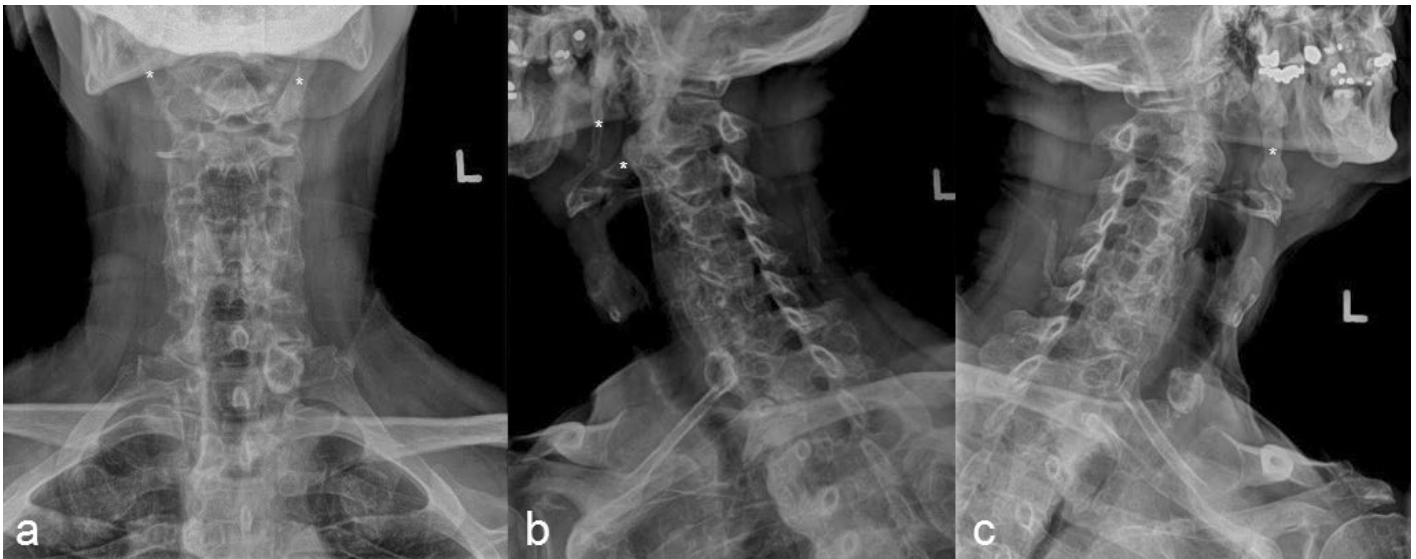


Figure 3. Anteroposterior and oblique cervical spine radiographs also reveal the ossified stylohyoid ligaments. The right stylohyoid ligament appears bipartite with the apparent formation of a central pseudoarticulation dividing it.

oid ligaments can be palpated intraorally along the tonsillar pillars and radiographic imaging usually confirms the diagnosis.¹⁻³ Anatomically, these bony structures can compress or irritate the surrounding neurovascular soft-tissues in the neck, namely the internal carotid artery and the trigeminal, facial, glossopharyngeal, and vagus nerves. Patients may present with an array of symptoms although many with elongated styloid processes or ossified stylohyoid ligaments will also remain asymptomatic.^{3,4} Table 1 lists the key imaging and clinical features for Eagle syndrome and Table 2 lists the key imaging features of DISH. Symptomatic patients with Eagle syndrome can be treated surgically or non-surgically.^{3,5,6}

Table 2.
Key imaging features of DISH

Key imaging features
<ul style="list-style-type: none"> • Flowing hyperostosis • Preservation of disc spaces • Preservation of facet joints (no arthrosis or ankylosis) • Displacement or compression of prevertebral soft-tissues and airway

Chiropractors and other manual therapy providers should note, however, that elongated styloid processes or ossified stylohyoid ligaments may pose a relative contraindication to thrust manipulation of the cervical spine.⁷ For instance, an association between styloid process length and risk of cervical carotid artery dissection has been reported.⁸ Surgical treatment for Eagle syndrome typically involves styloid process shortening or resection.^{3,5}

The etiology of Eagle syndrome is still controversial but it is thought to result from post-surgical (e.g. tonsillectomy) or traumatic (e.g. styloid fracture) scarring.¹⁻³ There is some evidence to suggest that there may also be a correlation between ossification of the stylohyoid ligaments and ligamentous ossification of the cervical spine in patients with DISH.⁴ This combination of findings is unique, however, in that very few cases involving both Eagle syndrome and DISH have been described in the literature.^{4,9} As such, further investigation is needed in order to validate this association.

In the current case, the patient consulted his primary care physician and no further diagnostic testing or surgical treatment was recommended. A course of conservative care, including neck mobilizations (i.e. cervical flexion-distraction / decompression therapy) and instru-

ment-assisted soft-tissue massage (i.e. Graston® Technique), was implemented. The soft-tissues treated included the cervical paraspinals, scalenes, upper trapezius, and levator scapulae muscles, bilaterally. After seven visits (over 10 weeks), the patient reported a mild decrease in neck pain and stiffness but continued to have symptoms of “choking” and asphyxia, particularly with neck flexion. It is unknown if these symptoms were as a result of Eagle syndrome, DISH, or a combination of both. Several cases of dysphagia and airway obstruction in relation to DISH and/or Eagle syndrome have been reported.^{9,10} In the current case, an upper G-I fluoroscopic (barium swallow) study could have been performed in order to more definitively ascertain the cause of the patient’s oropharyngeal symptoms. Regardless, the clinical findings of neck pain, throat irritation, mild dysphagia, and ossified stylohyoid ligaments were compatible with a diagnosis of Eagle syndrome. For more information and additional examples of Eagle syndrome, visit Radiopaedia.org.¹¹

Key Messages

- Published cases involving Eagle syndrome and DISH of the cervical spine are rare
- The differential diagnosis of patients who present with dysphagia, a sensation of a foreign body in the throat, pain in the distribution of the carotid arteries, and/or neuralgia involving cranial nerves 5, 7, 9, and 10 should include Eagle syndrome
- Patients with this disorder can be treated surgically or non-surgically, however there is a relative contraindication to thrust manipulation of the cervical spine

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