CLINICAL VIGNETTE

An Unexpected Cause of Neck Pain and Swelling

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Introduction

The differential for neck swelling and pain is broad. We present below an uncommon cause of neck pain in a patient with underlying rheumatic disease.

Case

A 44-year old female with recently diagnosed Sjogren's Syndrome was seen for chronic, intermittent neck swelling, left greater than right.

She described chronic sensation of neck swelling for months, starting below the ear and extending down the neck on either side. Swelling was sometimes associated with discomfort with head flexion, and a full sensation in her throat. It was otherwise not painful, with no associated rash, fever, malaise, numbness or weakness. On examination, eyes and mouth were dry, with no lacrimal, parotid or submandibular hypertrophy. There was a subtle fullness noted along the bilateral neck. There was no palpable lymphadenopathy. Labs demonstrated high titer SSA (84 units) and positive ANA (>1:1280 homogenous). Inflammation markers at onset of symptoms were low. She had normal complete blood count and serum protein electrophoresis. A cervical x-ray demonstrated vertebral osteophytes between C4-C6 without soft tissue calcifications. Neck ultrasound demonstrated no cervical lymph node enlargement. The thyroid was homogenous, without thyroid or parathyroid nodules. Non-contrast computed tomography of the neck demonstrated locules of air pockets along the right paratracheal and para-esophageal region, consistent with likely tracheal diverticulum, and calcified stylohyoid ligaments bilaterally. There was ossification of the posterior longitudinal ligament at C5 and C6. There were calcifications in the right palatine tonsil, and normal appearance of submandibular glands. No pathologic cervical lymphadenopathy was noted. Esophagus barium swallow and esophagogastroduodenoscopy were normal.

Given stylohyoid ligament calcification, Eagle Syndrome was considered. She was referred to her pulmonologist and oto-laryngologist regarding suspected tracheal diverticulum. Her, neck swelling and discomfort resolved with self-massage, without further intervention.

Discussion

Eagle syndrome is named after Watt W. Eagle, who reported an elongated styloid process as an uncommon etiology of facial pain and headache in the 1930s.[1] Symptoms can also include dysphagia, odynophagia, globus sensation, otalgia and tinnitus. The stylohyoid process length can vary, with the normal range reported as 2.5-3 cm. The stylohyoid ligament extends from the styloid process to the hyoid bone, and functions to suspend the hyoid bone. Anatomically, the stylohyoid process and apparatus are in close proximity to the internal carotid artery, internal jugular vein, accessory, hypoglossal, vagus and glossopharyngeal nerves.[2] An elongated stylohyloid process can be seen incidentally in about 4-30% of the general population, although only less than 5% may have symptomatic disease. There is a male/female ratio of 1:3, and presentation can be unilateral or bilateral.[3,4]

Two forms of Eagle syndrome were initially described, relating to "classic" (pain along cranial nerve distribution as above, post-tonsillectomy or trauma) and "stylo-carotid-artery" (vascular compression, carotidynia) presentations. The vascular type Eagle syndrome has been reported as a rare cause of internal carotid artery dissection and stroke.

Our patient had near complete calcification of the bilateral stylohyloid ligaments and typical neck pain consistent with Eagle syndrome. Calcification was not reported on initial cervical radiographs, although visible on these films (Figure 1). They were better visualized on computed tomography of the neck. Interestingly, prior imaging noted pelvis enthesopathy and ossification of the posterior longitudinal ligament. Stylohyloid ligament calcification has been postulated to be a manifestation of enthesopathy.[5,6] Trauma, endocrine abnormalities and calcium imbalance have also been discussed.[7]

Given the patient's history of Sjogren's Syndrome, alternate etiologies of neck swelling, including lymphadenopathy, infection and salivary gland enlargement were initially suspected. Fortunately, imaging and lab work were not supportive. We also discussed large cervical osteophytes as an uncommon cause of dysphagia, although this did not fully explain her symptoms. Tendinosis at the stylohyoid insertion can also mimic symptoms of Eagle syndrome.

Management options for Eagle Syndrome include conservative treatment with analgesics and nonsteroidal anti-inflammatory drugs, local steroid or anesthetic injections, and for recalcitrant disease, styloidectomy.

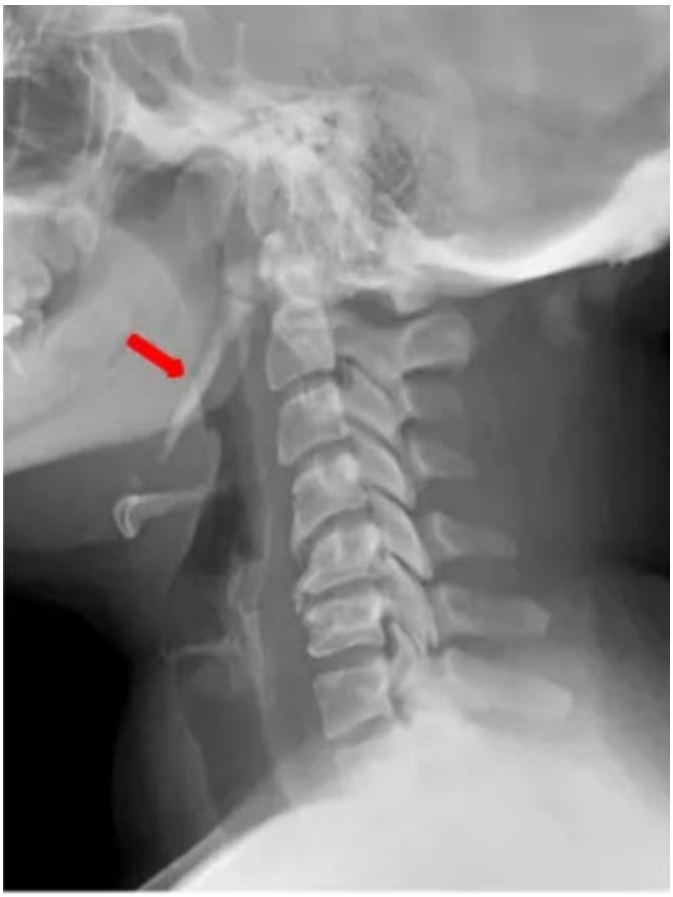

Figure 1. Radiograph of the cervical spine, lateral view, demonstrating ossification of the stylohyoid ligaments (red arrow), and C4-6 osteophytes.

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