

JCHS-IQ-01-2017

ANSWER TO JCHS-IQ-01-2017

Eagle's syndrome

CT of the neck demonstrated bilateral proximal stylohyoid ligament ossification. Bilateral styloid processes are normal. (Figures 1 and 2). The oropharynx, nasopharynx and hypopharynx are grossly normal. The findings are in keeping with Eagle's syndrome.

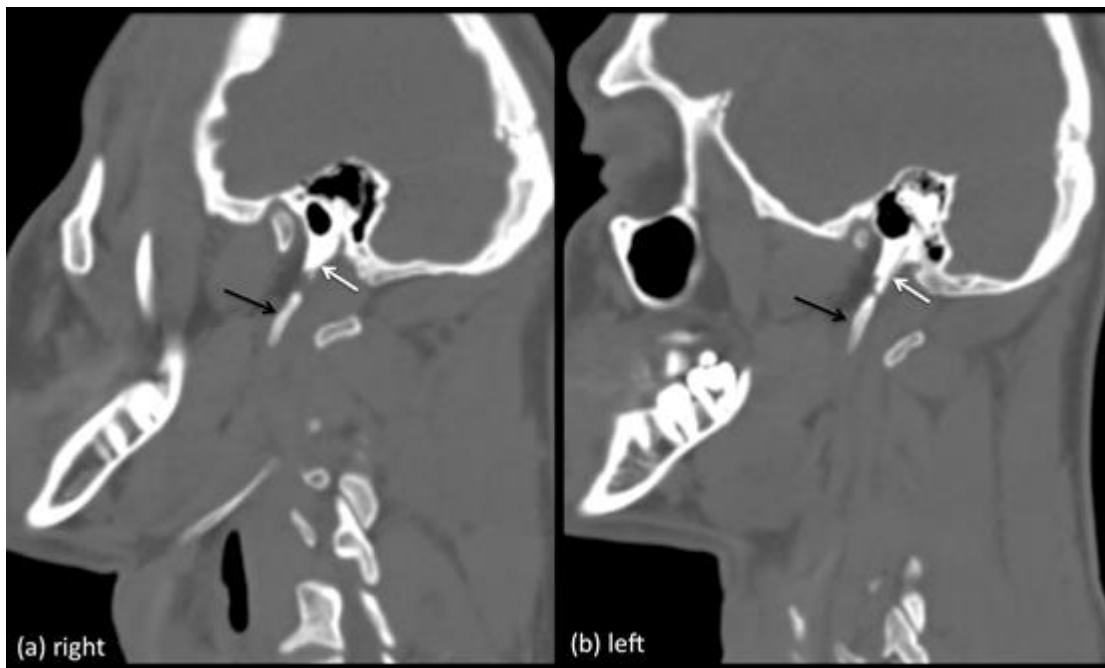


Figure 1 CT sagittal oblique multiplanar reconstruction of the neck demonstrated ossified stylohyoid ligament measuring 1.7cm in length on the left and 1.6cm on the right (black arrow). It extended to the level of C1/C2. The normal styloid process is identified and measures 1.2cm each (white arrow)

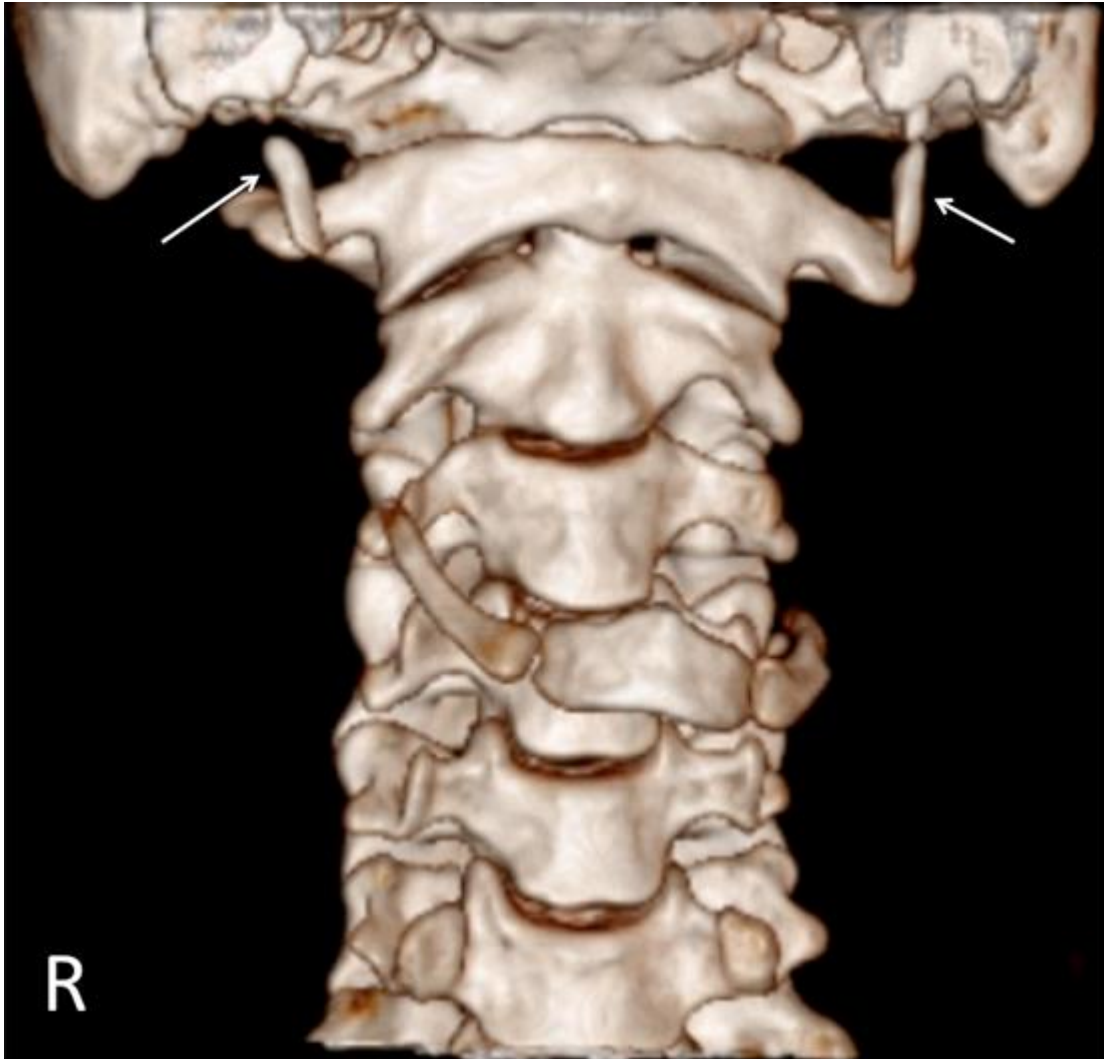


Figure 2 Volume Rendering Technique (VRT) reconstruction of the CT demonstrated bilateral ossification of the stylohyoid ligament (white arrows)

Eagle's syndrome was first described in 1937 by an otolaryngologist at Duke University, Watt W. Eagle (1). It can present with a multitude of symptoms caused by elongated styloid process or calcified stylohyoid ligament. Patients can present with otalgia, dysphagia, foreign body sensation in the throat or impingement of the carotid artery causing headache (2). Although 4% of the population may have an elongated styloid process, only a small percentage (4% to 10.3%) of this group may actually be symptomatic, giving a true incidence of about 0.16% (3). It is usually bilateral. The pathogenesis of this disease is unclear, although several theories have been suggested (3, 4):

- Persistence of embryologic precursors of the styloid resulting in a congenital elongation of the styloid process
- Persistence of cartilaginous component at the insertion of the stylohyoid ligament which then becomes ossified

Diagnosis is made clinically and further confirmed by imaging. A palpable styloid process which produces symptoms upon compression suggests the diagnosis. However, there are asymptomatic patients with palpable styloid process, making it difficult to establish a causal relationship between the disease and symptoms. Initial imaging by lateral X-ray of the neck may demonstrate the elongated styloid. Further evaluation by CT is usually indicated to confirm the diagnosis. CT can also demonstrate the relationship between the styloid process and the surrounding soft tissue, such as possible compression of the carotid artery or oropharynx. Exclusion of other diseases that may cause similar symptoms can also be made with CT. With technological advances, Volume Rendering Technique (VRT)

reconstruction of the CT can produce a 3D visualisation of the elongated styloid which will enable surgeons to accurately measure and plan further intervention. Treatment is both surgical (excision of the styloid process) or non-surgical (reassurance, nonsteroidal anti-inflammatory medications or steroid injections).

Learning Points

- Although uncommon, Eagle's syndrome is known to cause chronic otalgia despite multiple medications.
- It arises from either calcified stylohyoid ligaments or elongated styloid process and may also cause dysphagia, foreign body sensation in the throat or headache
- Disease awareness allows investigation by appropriate imaging such as CT to aid diagnosis and its subsequent management.

REFERENCES

1. Eagle W. Elongated styloid process: report of two cases. *Arch Otolaryngol.* 1937; 25(5): 584-7.
2. Fini G, Gasparini G, Filippini F, Becelli R, Marcotullio D. The long styloid process syndrome or Eagle's syndrome. *J Craniomaxillofac Surg.* 2000; 28(2): 123-7.
3. Murtagh RD, Caracciolo JT, Fernandez G. CT findings associated with Eagle Syndrome. *AJNR Am J Neuroradiol.* 2001; 22(7): 1401-2.
4. Bafaqeeh SA. Eagle syndrome: classic and carotid artery types. *Otolaryngol Head Neck Surg.* 2000; 29(2): 88-94.

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