

ELONGATED STYLOID PROCESS (EAGLE'S SYNDROME) & SEVERE HEADACHE

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ABSTRACT : Elongated styloid process as cause of throat pain & otalgia is common, as found in classical Eagle's Syndrome. However, elongated styloid presenting with headache due to impingement on carotid artery and neurological structures like glossopharyngeal nerve, is very uncommon presentation. Here such a case with fifteen years of history of being treated as migraine, relieved by styloidectomy by Eagle's technique and with a long symptom-free follow up of several years, is presented. For the clinician it is important to be aware of such clinical variants to successfully diagnose such head & neck pain cases, as resecting them surgically, resolves the symptoms completely.

Key words : Elongated styloid process (ESP), Eagle's Syndrome., Headache., Carotid Artery impingement., Glossopharyngeal nerve impingement.

INTRODUCTION

Normal length of styloid is 2cm to 3cm in adults and longer than 3cm is found in 4-7% of the population. Radiological evidence of elongated styloid process found in some individuals, who are symptom free, is of no clinical significance and can be accepted as normal anatomical variants. Classically, an elongated styloid process or calcified stylohyoid ligament causes Eagle's syndrome which was originally described by Watt W. Eagle in 1937. However, certain number of such patients may not have cervicofacial complaints of Eagle's syndrome and they may present with uncommon symptoms mimicking neurological or infectious diseases (Yetiser et al, 1997). Symptoms related to Eagle's syndrome can be conferred with those attributed to a wide variety of facial neuralgias, migraine headaches and/or oral, dental or temporo-mandibular joint disorders.

Carmada et al (1989) proposed that symptoms similar to stylohyoid syndrome can occur, in not a true calcified stylohyoid ligament (absence of radiographic evidence of ossification) & they are termed as 'Pseudo-Stylohyoid Syndrome'. A case of elongated styloid process with severe throbbing headache is presented here.

CASE REPORT

M.K. 38 years old lady presented with foreign body sensation, itching pain in right ear, right sided facial swelling, neck pain, and severe headache for 5 years. Foreign body sensation of ear used to be relieved momentarily with neck rotation. Ear pain used to increase with chewing of hard eatables, drinking of hot tea and yawning. Occasional mild to moderate headache was complained since 1977. However, for last 5 years it was severe, continuous, throbbing. It was usually in

both temples (more with chewing hard eatables) and pronounced on right side of head, but the entire head was painful at times. Insomnia was a frequent feature and sometimes patient had to literally bang her head on the walls, pain killers and sleep inducing injections were not effective. For last 15 years, this headache was treated as migraine by drugs like vasograin. Tegretol was used for sometime but discontinued due to dizziness.

On examination - Nose & tympanic membranes were normal. In the throat-right tonsil was protruding more, bony hard styloids were palpable on both sides, right side's tip projecting in lower tonsillar fossa and on palpation the patient experienced severe head pain of the type she had complained of. There was also tenderness between upper end of sternocleidomastoid muscle and angle of mandible & also along the carotid artery. Her ECG, X-ray of chest, sinus and mastoids, blood sugar and thyroid function tests done earlier were normal. Skull antero-posterior radiograph (open mouth) showed markedly enlarged, curved and thick right styloid. Left styloid was also markedly enlarged but less dense. Patient was taken for surgery by intra-oral route (Eagle's technique) – right styloid process was very curved and protruding deeply into lower tonsillar fossa. On the right side 3.5cm long and on the left side 2.8cm long styloid processes were removed. Severity of headache, FB ear sensation immediately disappeared. Mild aching ear and head pain persisted for 1½ months which was taken care of by Tegretol. After this patient became symptom free, without use of painkillers. Her follow-up, (6 years after operation), except for occasional headache showed absence of regular head pains and sleep disorders. But her right sided mild and occasional neck pain was diagnosed as cervical

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Fig. I : Skull A.P. Radiograph showing bilateral elongated styloid process.
Rt. Thick, curved.

spondylosis which got better with cervical exercises and other treatment.

Embryologically, there are four segments of the stylohyoid complex. These template derived from Reicher's cartilage are the styloid process, stylohyoid ligament, lesser cornu of hyoid and superior portion of hyoid corpus. In lower species, there can be a chain of four bones performing function of the stylohyoid ligament. Remains of cartilage in styloid ligament has a potential to mature in normal bone and that is how the ligament ossifies. It is the ossified ligament which is known as the elongated styloid process. Ossification of ligament can occur anywhere along its length, occasionally with segmentation and pseudo-articulation. In addition to the calcification theory, congenital elongation of styloid process due to persistence of a cartilaginous anlage in the stylohyale and growth of osseous tissue at insertion of the styloid ligament, are another etiology of enlargement. Carmada et al (1989) proposed a fourth theory to explain the presence of symptoms in absence of radiological evidence of ossification. With aging process, soft tissue loses elasticity, a localized inflammatory reaction causes tendonitis at junction of ligament and lesser cornu. Ligament stiffens and inflammatory response of associated tissue causes the symptoms. This would not be a true calcified ligament and is known as pseudo-stylohyoid syndrome.

Stylohyoid ligament arises from the tip of styloid process to insert into the lesser cornu of hyoid directly by a tendinous insertion, instead of connection with periosteum (Kreps YP

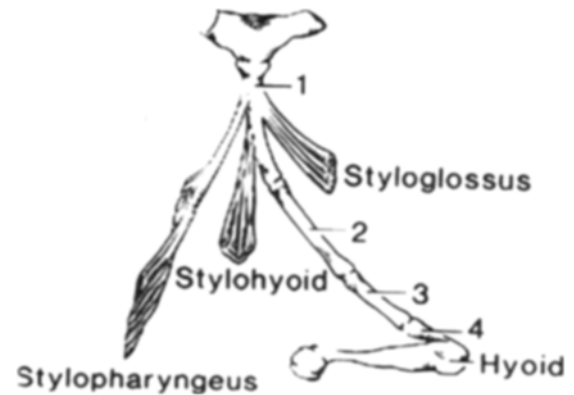


Fig. II : Showing parts of stylohyoid complex (1) Styloid (2) Styloid ligament (3) Its tendon (4) Lesser cornu of hyoid.

et al, 1981). Elongated styloid process may sometimes get displaced from normal position. Radiologically elongated ossified styloid process may be smooth and well corticated or bulky and irregular.

Symptoms of Eagle's syndrome are thought to be due to relationship of abnormal stylohyoid ligament to neighbouring anatomic structures. The important muscular, neural and vascular structures include medially, internal jugular vein with accessory, hypoglossal, vagus and glossopharyngeal nerves; internal carotid artery and superior constrictor muscle (near tip); lateral to the tip of process, is the external carotid artery, which bifurcates into superficial temporal and maxillary arteries.

Several patho-physiological mechanisms leading to pain have been proposed : (1) Traumatic fracture of the styloid process causing proliferation of granulation tissue, which may cause pressure on the surrounding structures. (2) Compression of the adjacent nerves, the glossopharyngeal, lower branch of the trigeminal or chorda tympani. (3) Degenerative & inflammatory changes in tendonous portion of the styloid insertion, called insertion tendonosis. (4) Irritation of the pharyngeal mucosa by direct compression or post tonsillectomy scarring (involves cranial nerves V, VII, IX & X) and (5) Impingement on carotid vessels producing irritation of the sympathetic nerves in the arterial sheath.

Eagle's syndrome involves a pattern, of head pain due to elongation of styloid process. Two major patterns occur. Eagle (1958) described, the classical syndrome as persistent pain in pharynx, aggravated by swallowing, with pain

frequently referred to the ear on the same side. He also noted increased salivation, hesitancy and difficulty of swallowing, gagging and a foreign body sensation. Styloid impingement is usually on glossopharyngeal nerve (main basis of neurological symptoms of Eagle's syndrome) as it is in close proximity to styloid ligament and related muscles but other cranial nerves may be affected. He attributed the pain due to the scarring around styloid tip after tonsillectomy. Pharynx pain may be sharp but less severe than lancinating pain of glossopharyngeal neuralgia. The symptoms of elongated styloid process are increased due to its attachment, by breathing, yawning, hyperextension of neck besides swallowing.

The second symptom category described by Eagle was the styloid process-carotid artery syndrome which could occur regardless of whether a tonsillectomy had been done. He theorized that styloid process impinged on carotid artery on affected side causing carotodynia, intermittent frontal, parietal or temporal headache, otalgia and dizzy or blackout spells. He noted that these patients had tenderness on palpation of carotid artery. Impingement against internal carotid artery causes parietal headache or head-pains throughout the area supplied by ophthalmic artery with little or no pain below the eye. If the external carotid artery is involved, patient complains of homolateral face pains which occur below the eye. (Ryan, 1978).

In this patient, right sided headache all over is possibly explained by compression of both carotid arteries along with neurological impingement. It appears that markedly enlarged, thick and curved styloid is more important in causing severe symptoms.

Koebke (1976) reported a 64 years old male whose left carotid artery is compressed by anomalous styloid process and by the tendon of stylohyoid muscle. Such cases are reported in literature. Histological examination of the wall at the level of compression, revealed early arteriosclerosis. Massey EW & J. (1979) also reported elongated styloid process as cause of hemicrania. Monks FT (1978) reported styloid as probable cause of facial pain. Montalbetti et al (1995) reviewed Eagle's syndrome and concluded that elongated styloid process is the cause in first classical group characterized by unilateral dysphagia and pharyngeal pain radiating to the ear and worsened by swallowing and in those a typical painful head

& neck conditions related to elongated styloid process and relieved by styloidectomy. They stressed that this syndrome deserves consideration in International headache classification.

DISCUSSION

Embryologically, there are four segments of the stylohyoid complex. These template derived from Reicher's cartilage are the styloid process, stylohyoid ligament, lesser cornu of hyoid and superior portion of hyoid corpus. In lower species, there can be a chain of four bones performing function of the stylohyoid ligament. Remains of caretohyal cartilage in styloid ligament has a potential to mature in normal bone and that is how the ligament ossifies. It is the ossified ligament which is known as elongated styloid process. Ossification of ligament can occur anywhere along its length, occasionally with segmentation and pseudo-articulation. In addition to calcification theory, congenital elongation of styloid process due to persistence of a cartilaginous anlage in the stylohyale and growth of osseous tissue at insertion of the styloid ligament, are another etiology of enlargement. Carmada et al (1989) proposed a fourth theory to explain the presence of symptoms in absence of radiological evidence of ossification. With aging process, soft tissue loses elasticity, a localized inflammatory reaction causes tendonitis at junction of ligament and lesser cornu. Ligament stiffens and inflammatory response of the associated tissue causes the symptoms. This would not be a true calcified ligament and is known as pseudo-stylohyoid syndrome.

CONCLUSION

It is important for clinicians to be aware of clinical variants of elongated styloid process with a typical symptom or to seek origin of head-neck pain before reaching any conclusion. Elongated styloid process may give rise to pain in head, cheek, chin, neck along distribution of carotid artery. A careful history, intra-oral digital palpation of tonsillar fossa and exacerbation of pain (similar to those complained by this patient) by palpation, carotid artery tenderness (increased by palpation and head movement) and hyoid tenderness, standard skull radiograph, and relief of symptoms by surgery are all confirmatory of the diagnosis (ruling out other differential diagnosis of migraine like headaches, neuralgias, a typical face pain etc.). A high index of suspicion should be maintained in all cases of head, face and neck pains and a proper otolaryngologic examination should be a routine in such cases.

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