Eagle's syndrome with 3-D Reconstructed CT: two cases report

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Eagle's syndrome is a rare disease which usually occurs in adult patients aged 30 to 50 years. It is characterized by the symptomatic elongation of the styloid process or mineralization of the stylohyoid ligament complex. The symptoms ranges from mild discomfort to acute neurologic and referred pain. Clinical diagnosis is based on palpating the tonsillary fossa, which should reveal a bony formation and exacerbate pain. Confirmations can be made by a lateral x-ray film or panorex radiograph. CT and 3-D reformatted technique provided more information for surgeons. We reported two cases of Eagle's syndrome and evaluated by 3-D reformatted CT.

Key words: Computed tomography(CT), three dimensional; Eagle's syndrome

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Eagle's syndrome is characterized by the symptomatic elongation of the styloid process or mineralization of the stylohyoid ligament complex. The symptoms ranges from mild discomfort to acute neurologic and referred pain. A few cases had been reported in Chinese people. Confirmations can be made by radiographic studies. We reported two cases of Eagle's syndrome and evaluated by 3-D reconstructed CT.

CASE REPORT

Case1

A 49 year-old man with unremarkable medical history but complained of intermittent odynophagia and lumping throat sensation for 6 months. The pain was more severe on the left side and aggravated by swallowing. A tender point at left tonsil was noted during physical examination. Neck CT with 3-D reformation showed elongated ossified left styloid process (Fig. 1-2). Eagle's syndrome was diagnosed. The injected left tonsil and 3.0 cm of the left styloid process was excised through transpharyngeal approach. Patient was free of symptom after the operation.

Case2

A 55 year-old man presented with a hisotry of rectal cancer with liver metastasis. He suffered from dysphagia, sorethroat, headache, voice change and hypersalivation. The throat pain was exacerbated with head rotation. Neck CT was performed for suspected metastatic lesions. It showed marked elongation of bilateral styloid processes with ossification of bilateral stylohyoid ligaments and thyroid cartilage (Fig. 3). Plain film of cervical spine revealed typical appearance of Eagle's syndrome (Fig. 4). No operation was arranged due to his poor condition.

DISCUSSION

Eagle's syndrome is a rare disease entity characterized by the symptomatic elongation of the styloid process or mineralization of the stylohyoid ligament complex [1]. The anatomic anomaly was first mentioned by Marchetti in 1652 and sporadically reported in the nineteenth century. Eagle described the syndrome in 1937, identifying two forms of this disease based on the symptoms of his patients [2, 3]. These patients were categorized into two groups: those who had classical symptoms of a "foreign body" lodged in the throat with a palpable mass in the tonsillar region following tonsillectomy; and those with pain in the neck following the carotid artery distribution (carotid artery syndrome) [4, 5].

The epidemiological incidence of Eagle's syndrome is variable in the literature [3]. It occurs usually in adult patients aged 30 to 50 years, but a few suspicious cases in children have been reported [6]. Keur et al. described a greater incidence of elongated styloid process in the higher age groups. Symptoms are more common in females. No statistically significant sex-specific difference was found [7].

There is no obvious correlation between the severity of pain and the extent of ossification of the stylohyoid complex. Many patients with incidental finding of an elongated styloid process are asymptomatic [8]. Variable symptoms presented are due to the complicated anatomical relationships of styloid process. It has attachment to the stylopharyngeal, stylohyoid, and styloglossal muscles, and medially the superior pharyngeal constrictor muscle, pharyngobasilar fascia, internal jugular vein, and the accessory, hypoglossal, vagus, and glossopharyngeal nerve. The stylohyoid ligament connects the styloid process with the small cornus of the hyoid bone [9].

The symptoms range from mild discomfort to acute neurologic and referred pain, including continuous pain in the throat, sensation of a foreign body in the pharynx, difficulty in swallowing, otalgia, headache, pain along the distribution of the external and internal carotid arteries, dysphagia, pain on cervical rotation, facial pain, vertigo, and syncope [10, 11]. Clinical diagnosis is based on palpating the tonsillary fossa, which should reveal a bony formation and should exacerbate pain. The pain is also triggered by head rotation, lingual movement, swallowing, or chewing [9]. The differential diagnosis of pharyngeal pain with radiation to the ear, face, or neck is broad [12]. Head and neck tumors, cranial nerve neuralgias, pharyngotonsillitis, or temporo-mandibular joint disease should be excluded.



Figure 1. Case 1: Non-contrast CT scan of neck reveals elongation of the stylohyoid ligaments (arrows).



Figure 2. Case 1: Neck CT with 3-D reconstruction reveals elongation and fracture of the left stylohyoid ligament (arrow).

Eagle reported the normal styloid process is approximately 2.5 to 3.0 centimeters in length [4]. A 3-cm or longer process is considered anomalous [10, 16]. The symptoms were initially attributed by Eagle to scarring around the styloid tip following tonsillectomy in recently operated patients [9]. But the majority of symptomatic patients have had no recent history of tonsillectomy or other cervicopharyngeal trauma [8]. Some theories have been proposed: 1. Congenital elongation of the styloid process due to persistence of a cartilaginous analog of the stylohyal (one of the embryologic precursors of the styloid), 2. Calcification of the stylohyoid ligament by unknown process, and 3. Growth of osseous tissue at the insertion of the stylohyoid ligament [12].

If highly suspicious for Eagle syndrome, confirmation can be made by radiographic studies. A lateral



Figure 3. Case 2: Neck CT with 3-D reconstruction showed ossification and elongation of bilateral stylohyoid ligament and thyroid cartilage (arrows).



Figure 4. Case 2: Plain film of cervical spine showed ossification of bilateral stylohyoid ligament and thyroid cartilage (arrows).

sliced x-ray film or panorex radiograph usually shows an elongated styloid process. CT provides complementary information to that provided by plain radiographic studies [13]. Reconstructed 3-D technique is useful for the diagnosis and facilitates explanation to the patients [14,15]. Treatment of Eagle syndrome can be pharmacologically or surgically. Pharmacological treatments include reassurance, nonsteroidal anti-inflammatory medications, and steroid injections in the anterior pillar of the tonsillary fossa [16]. There are two methods for surgical excision of the styloid process and/or the mineralized ligament [1]. Transpharyngeal approach was used for one of our cases. It has better cosmetic effects. An external approach is easier to perform and reduces hemorrhagic and cervical infection, but results in a cutaneous scar [17,18].

Eagle's syndrome, though not a common entity, is probably underdiagnosed. Surgeons should be aware of the clinical condition and arrange proper image studies. Although Eagle's syndrome can be confirmed by plain films, CT is more useful to demonstrate the location and extent of the elongated styloid process. Utilizing the image process technique, reconstructions in other dimensions and 3D image yields additional information and enables an anatomic study of the adjacent structures.

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Eagle症候群的三度空間電腦斷層重組影像: 雨病例報告

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Eagle症候群是一種少見的疾病,好發在三十至五十歲的成年人。主要是莖突過長或莖狀舌 韌帶之骨化引起的症狀。表現可以從輕微不適到急性神經症狀。臨床上是以扁桃腺窩觸摸到骨 質硬塊並引發疼痛來診斷。藉由側位X光片或Panorex片來確診。電腦斷層及三度空間影像重 組可以提供外科醫師更多的資訊。我們報告兩例以三度空間電腦斷層做評估的Eagle症候群。

關鍵詞:電腦斷層,三度空間重組;Eagle 症候群