

A Rare Cause of Recurrent Cerebral Ischemia and Syncope: Eagle Syndrome

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Abstract: Eagle syndrome is defined as symptomatic elongation of the styloid process or calcification of the stylohyoid and stilo-mandibular ligament. The syndrome was described by WW Eagle in 1937. The styloid process is located between the internal and external carotid arteries and laterally in the tonsillar fossa. Patients with cerebrovascular ischemia causing syncope or hemiparesis due to Eagle syndrome are rarely published in the literature. The authors presented a patient with recurrent cerebrovascular attacks due to long styloid process.

Key Words: Cerebral ischemia, Eagle syndrome, recurrent embolism, stylocarotid syndrome, surgical treatment

Eagle syndrome (ES) is defined as the symptomatic elongation of the styloid process or calcification of the stylohyoid and stilo-mandibular ligament.¹ The syndrome was described by WW Eagle in 1937.² The styloid process is located between the internal and external carotid arteries and laterally in the tonsillar fossa.³ Symptoms related to ES are nonspecific including recurrent throat pain, foreign-body sensation in the pharynx, limited mouth opening, trismus, tinnitus, and cervical pain.³ Patients with cerebrovascular ischemia due to ES are rarely published in the literature. We presented a patient of recurrent cerebral ischemia due to long styloid process with the related recent literature.

CLINICAL REPORT

A 33-year-old male patient with a history of recurrent cerebrovascular event was consulted from neurology department for the treatment of long styloid process. He had no tonsillectomy history. Bilateral ES was diagnosed when the patient complained of recurrent episodes of embolism, hand-foot weakness, and syncope. Selective cerebral angiography examination and three-dimensional (3D) computerized tomography images demonstrated bilateral internal carotid artery dissection and bilateral ES (Fig. 1A). Bilateral internal carotid artery dissection and bilateral internal carotid stenosis were observed in the localizations matching the styloid process in the angiography of the patient. On axial computerized tomography, the right styloid process was found to be 42 mm and the left styloid process was found to be 41 mm (Fig. 1B). Bilateral styloid process

excision using bone scissor with transcervical approach was performed under general anesthesia (Fig. 1C). Intraoperative and postoperative complications were not seen. The patient was discharged on the second postoperative day after the treatment. No neurological symptoms were seen in postoperative 1-year follow-up visits.

DISCUSSION

Long styloid process or ES is mostly diagnosed for symptomatic patients with a styloid process longer than 3 cm on the panoramic radiographs or computerized tomography scans.⁴ The underlying pathogenesis of ES is not fully understood. Congenital elongation of the styloid process, calcification of the stylohyoid ligament, formation of osseous tissue at the stylohyoid ligament site, and fibrosis due to tonsillectomy or pharyngeal surgery were thought to be possible etiological factors.^{2,5} Two different clinical forms have been identified in ES. Classical form occurs due to surgical trauma after tonsillectomy. The scar after tonsillectomy causes contracture in the tonsil area and thus causes tension on the long styloid process. Tonsillar fossa pain, foreign-body sensation, reflected ear pain can be seen. These complaints occur due to the irritation of the 5th, 7th, 9th, and 10th cranial nerves.²

The second clinical form is the stylocarotid syndrome. This clinical form is not related to the previous pharyngeal surgery. In this form, central nervous system complications occur due to mechanical irritation of the vascular walls of the internal and external carotid artery by the elongated styloid process. Cerebrovascular manifestations of ES can reflect a wide range of mechanism including cerebral ischemia from vascular compression, injury to the carotid arteries with embolization or dissection resulting in transient ischemic attack or hemiplegia, and vasovagal response.^{5,6} Our patient had bilateral internal carotid artery dissection. Symptoms can be various including aphasia, visual symptoms, weakness, and syncope.^{5,6} Although conventional radiographs and panoramic radiographs can be used for diagnosis of ES,⁷ 3D computerized tomography scans are useful for demonstrating the topographic anatomic relationship between styloid process and neighboring neurovascular structures.^{3,4,8,9} Due to the great variations in the length, angulation and

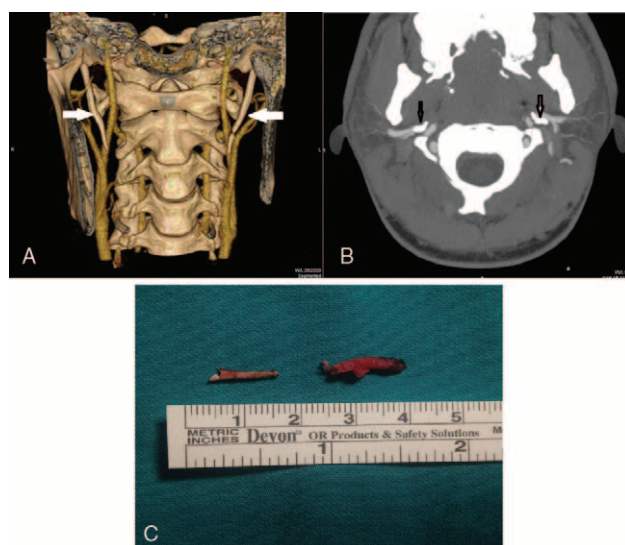


FIGURE 1. (A) Three-dimensional computerized tomography image. (B) Computerized tomography axial image. (C) Bilateral styloid process excision materials.

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proximity of styloid process to internal and external carotid arteries,⁸ 3D computed tomography scans are useful diagnostic tools that can present the compression effect of styloid process to carotid arteries to rule out other reasons of cerebrovascular ischemia.^{5,10} We also performed 3D computed tomography scans for our patient. Although medical treatment options such as nonsteroidal anti-inflammatory drug use, transpharyngeal steroid, or local anesthetic injection can be used for the treatment of ES, surgery is the main treatment choice for cases with cerebrovascular complications.^{5,6} For surgical option, intraoral and external approaches exist. In the intraoral approach, the operation time is shorter and there is no cervical scar tissue formation. Disadvantages include a narrow field of view and a risk of deep neck infection after surgery. Transcervical approach has the advantage of wide surgical field with dominating the important anatomical structures, whereas skin scar tissue formation is observed.¹¹ It is an effective and safe surgical option with limited morbidity.¹² Excision of the styloid process with Piezosurgery during external approach has also been advised.¹³ We also performed external approach for our case, but we preferred bone scissor.

CONCLUSION

Eagle syndrome should be kept in mind for patients with neurological symptoms such as recurrent syncope attacks and cerebrovascular ischemia. Three-dimensional computerized tomography images with angiography scans are the most appropriate preoperative diagnostic tools. Transcervical approach is the main surgical way to dominate the major vascular and neurological structures.

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