Eagle Syndrome and cervical disc herniation – combined symptomatology and treatment. Case report

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Abstract

Eagle syndrome is a rare condition where an abnormally elongated (over 3cm) calcified stylohyoid ligament irritates the adjacent soft tissues, causing a number of symptoms related to neck movements, such as local pain, odynophagia, dysphagia, otalgia and limited neck mobility. The symptoms depend from the structures involved: cranial nerves VII, IX, X and XII, as well as internal jugular vein and carotid artery. The conservative treatment of the condition includes: non-steroidal anti-inflammatory, antiepileptic drugs, physiotherapy and neck exercises, steroids, and local anesthetic injections. The possible surgical approaches are internal (transoral) or external – transcervical and parapharyngeal. The case is presented of a female patient who suffered neck and shoulder pain from the combined occurrence of left-sided elongated styloid and cervical disc herniation. She was successfully treated with resection of the styloid via parapharyngeal approach.

Key words

Eagle syndrome, cervical disc herniation, parapharyngeal surgical approach, neck pain

INTRODUCTION

Eagle syndrome is a rare congenital condition in which an abnormally elongated styloid process or calcified stylohyoid ligament irritate the adjacent soft tissues, causing a number of symptoms: local pain, odynophagia, dysphagia, hypertalvation, otalgia, limited neck mobility and transient visual loss (amaurosis fugax). Characteristically, the symptoms aggravate with changes in head position. The surrounding structures that can be involved are: cranial nerves VII, IX, X and XII, as well as internal jugular vein and carotid artery. Cases of carotid artery dissection or compression, causing recurrent TIA or ischemic stroke, have been reported [2]. Unless treated surgically with styloid process resection, the occlusion and damage to the crotid artery can be refractive to stenting [3]. In the case of conflict with the hypoglossal nerve, ipsilateral loss of tongue muscles and contralateral tongue deviation can be observed. The syndrome can be unilateral or, less frequently, bilateral. The imaging methods applied to diagnose the condition are: radiograph, orthopantogram and multi-slice 3D CT. The normal length of the process in adults is approximately 2.5 cm (ranging from 1.5–5 cm), a styloid over 3 cm is considered elongated [4, 5]. Some authors believe that the styloid is abnormal when its length exceeds one-third of the mandibular ramus in a panoramic view [6].

The syndrome was first described in 1937 by the American otorlaryngologist Watt Weems Eagle, who estimated the incidence of the elongated styloid in the general population to be 4% [7]. Eagle believed that the ossification of the styloid complex occurs after surgical trauma (tonsillectomy) or local chronic irritation [8]. The condition most frequently affects female patients in the 3rd – 5th decade of life [9]. In patients with temporomandibular disorder, the prevalence is reported to be as high as 76%, which seems to confirm the secondary to irritation character of the ossification[10]. A familial occurrence of Eagle’s syndrome was found and investigated by Morrison et al., who formulated the suggestion of an autosomal dominant heredity [11]. The authors point out that a similar condition – ossification of the posterior longitudinal ligament of spine (OPLLs) – prevalent in the Japanese population and causing cervical spine stenosis, has a traceable genetic etiology [12].

Palpation of the antero-lateral submandibular region can reveal the presence of an elongated styloid process which can be a trigger for the typical neuralgia [13]. Sometimes, the pathological mass can be palpated in the tonsillar fossa [14]. The differential diagnosis must consider: temporomandibular joint disease, migraine, headache, trigeminal, glossopharyngeal and sphenopalatine neuralgias [15].

The conservative treatment of the condition includes: non-steroidal anti-inflammatory drugs, physiotherapy and neck exercises, steroids, local anaesthetic injections. According to the authors, satisfactory pain control can be attained with pregabalin and amitryptiline [16]. Transpharyngeal, image-guided steroid and lignocaine injections do not provide sustainable pain-relief, the symptoms recurring after 6–12 months [17]. In most cases, lack of significant improvement leads to surgical treatment. The possible surgical approaches are internal (transoral) or external – transcervical and parapharyngeal. The first is retromolar and paratonsillar and requires less surgical and recovery time and leaves no visible scar. The latter allows wider visualization of the operative field and involves less risk of infection [18, 19]. A case series of 61 patients operated via submandibular external approach did not report any serious complications [20].
CASE REPORT

The patient, 54 year-old woman, a nurse, for five years had been having neck pain radiating to the left shoulder, and tingling and numbness of the left forearm and IV and V digits. In October 2014, the pain exacerbated, hand grip weakened and she began dropping objects from her left hand. She also noticed increasing stiffness of the neck and decreased mobility and pain as she tried to turn her head to the left. The pain spread to the left mandibular angle and left ear. The patient began suffering from odynophagia, vertigo and brief collapses. Additionally, she developed a hoarse voice.

The patient had been under close medical surveillance due to her previous neoplastic disease – in 2013 she had a hysterectomy because of menorrhagia, and histopathologic examination revealed a low grade endometrial cancer.

With the new symptoms she was referred to an otolaryngologist but neither the ear nor the parotid glands showed any abnormalities.

Cervical spine MRI and CT were performed and revealed cervical discs herniation – C5-C6 and C6-C7, as well as a calcified stylohyoid ligament. In the meantime, the patient consulted numerous specialists in different centers in Poland. A transoral approach to resect the enlarged styloid was proposed; however, the maxillofacial surgeon refused to perform the operation, anxious about the possible damage to the spinal cord caused by the positioning of the patient with concomitant cervical discopathy.

The patient was referred to a neurosurgeon for an elective surgical treatment. In February 2015, the patient underwent the surgery – open anterior C5-C6 and C6-C7 cervical discectomy with intervertebral cage stabilization. The skin incision was performed on the right half of the neck, according to the surgeon's preferences, enabling better exposure of the left nerve foramina. After completion of the discectomy, another incision – more cephalically and in the left submandibular area – was performed, the level confirmed by lateral x-ray with C-arm. The structures encountered and retracted in the dissection were: platysma, submental vein, mylohyoid muscle, stylohyoid and geniohyoid muscle, submaxillary gland and thyreohyoid muscle. The bony structure of calcified ligament was palpated and identified, then resected with a bone rongeur. Care was taken not to leave any sharp edges. After haemostasis was obtained, the wound was closed in layers; resorbable intracutaneus suture was used for skin closure. Good healing of the wounds followed, leaving minimal scars and a very satisfying cosmetic effect.

The symptoms of the patient resolved completely with no neurological deficits observed. The patient was discharged on the 3rd postoperative day and could restart her regular work after a month of rest.

REFERENCES


Figure 3. Resected specimen of the calcified stylohyoid ligament