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Case Report

A Novel Approach with Spinal Cord Stimulation for the Treatment of Chronic Pain in Eagle's Syndrome: A Case Report

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Abstract

Eagle's syndrome is a rare condition characterized by foreign body sensation in the throat, cervical pain, and dysphagia. It is commonly due to a pathological longer styloid process or ossification of the stylohyoid ligament. There are two classical therapeutic approaches: a conservative management, with a combination of Non-Steroidal Anti-Inflammatory Drugs (NSAIDs), anticonvulsants and antidepressants along with local injections of local anaesthetics/corticosteroids, or a surgical treatment i.e. the removal of the styloid process and stylohyoid ligament. We reported the case of a 52-years old woman presented with pain localized at the right side of the neck, back of the throat, tonsillar fossa, base of the tongue, soft palate and inferior part of the occipital bone together with sensation of foreign body in the throat. These symptoms started at the age of 29 after drinking some water with a pain intensity evaluated by numeric rating scale (NRS) of 10. Both pharmacological and surgical treatment didn't produce substantial relief. In 2017 the patient came to our attention and a 10KHz Spinal Cord Stimulator (SCS) was positioned between C2 and C7 with a significant pain relief (NRS4) and a complete relief from foreign body sensation in the throat. This is, to our knowledge, the first case of Eagle's syndrome treated with SCS. This case is here reported along with a short review of Eagle's syndrome and a description of mechanisms and potential applications of SCS.

Keywords: Chronic pain; Eagle's syndrome; High frequency spinal cord stimulation

Introduction

Eagle's syndrome, also known as styloid syndrome, was described for the first time by Watt W Eagle, an otolaryngologist of the Duke University in 1937 [1]. This syndrome is a rare condition where elongated temporal styloid processes, or calcified stylohyoid ligaments, are in conflict with the adjacent anatomical structures giving rise to a complex range of symptoms.

In the general population, the frequency of the elongated styloid process is estimated to be 4%, of which only 4% shows clinical manifestations [2,3]. The incidence of Eagle's syndrome is therefore estimated to be about 0.16% (1 in 62,500). Females have Eagle's syndrome about 3 times more often than males. Classic Eagle's syndrome is monolateral, however, rarely, it may be present on both sides. No significant difference is detectable between the right and left sides [4]. The syndrome is usually more frequent in young adults.

In terms of clinical expressions, are described: the "classic styloid syndrome" and the "second form". The first one is characterised by dull and persistent pain in oropharynx and face, dysphagia, and a foreign body sensation. The pain is actually centred in the tonsillar fossa that refers to the ipsilateral ear and get exacerbated by swallowing, yawning and chewing. Patient can also complain of dysphagia, Globus sensation and tinnitus can also occur. This form is typically seen in patients after pharyngeal trauma or tonsillectomy.

The "second form" of the syndrome ("stylocarotid syndrome") is characterized by the compression of the internal or external carotid artery (with their peri-vascular sympathetic fibres) by a laterally or medially deviated styloid process. Patient

complains of pain along the distribution of the artery, which is provoked and exacerbated by rotation and compression of the neck. It's not correlated with tonsillectomy. In case of impingement of the internal carotid artery, patients often refer supraorbital pain and parietal headache. In case of external carotid artery irritation, the pain radiates to the infraorbital region.

The literature contains many reports of transient ischemic attacks and stroke being associated with Eagle's syndrome. Moreover, elongated styloid process is a known risk factor for carotid artery dissection and also contains reports of dysphonia and glossodynia associated with an elongated styloid [5-7].

Eagle's syndrome can be diagnosed radiologically and by physical examination. The diagnostic golden standard is 3D CT reconstruction [8]. Sagittal CT angiography has a leading role in the radiological diagnosis of the stylocarotid syndrome and can provide further information regarding carotid flow especially if stroke or dissection is suspected.

Differential diagnosis requires the differentiation of the styloid syndrome from numerous cranio-facio-cervical painful syndromes. Scan is also helpful in deciding further management and guides the surgeon on how and from where to approach the surgery.

In both the classic and vascular form, the treatment consists in a conservative or surgical management. Conservative management is represented by the medical therapy which can be further divided into first-line analgesics such as Non-Steroidal Anti-Inflammatory drugs (NSAIDs) and second line consisting of a combination of anticonvulsants, antidepressants, local injection [9]. Also a stellate ganglion block added to medical therapy can result in near complete resolution of symptoms [10]. The most effective treatment of pain associated with Eagle's syndrome is the association of gabapentin, tramadol, and acetaminophen, associated with local injections of mepivacaine/dexamethasone [11-12]. The surgical approach turns out to be a definitive treatment and is associated with a better quality of life [13]. Surgical management can be divided into the intraoral and cervical approaches [14]. The traditional intraoral approach begins with a tonsillectomy, after identifying the tip of the styloid process. The dissection is brought to the periosteum of the styloid process and the ligaments are removed. The tip of the styloid is removed as proximally as possible and then the tonsillar fossa is sutured [13]. The complications related to this approach are infections and postoperative airway oedema [15]; moreover, in case of carotid lesion there is little control of bleeding. More recent intraoral techniques save tonsils.

The external cervical approach provides the best exposure; in fact, an incision is made at the level of the jaw and subperiosteal dissection of the styloid process is performed. Complication associated with this approach is the injury of the mandibular nerve [15].

Case Report

A 52-years old woman presented with pain localized at the right side of the neck, back of the throat, tonsillar fossa, base of the tongue, soft palate and inferior part of the occipital bone together with sensation of foreign body in the throat; these symptoms affected only the right side with a pain intensity evaluated with Numeric Rating Scale (NRS) of 10. Symptoms started at the age of 29 after drinking some water. Initially interpreted as a psychosomatic disorder, it was treated with anxiolytics and antidepressants. Over the years she underwent many therapeutic interventions (infiltration of local anaesthetics, sphenopalatine ganglion infiltration and radiofrequency, hypnosis, acupuncture) and pharmacological treatments (tramadol, pregabalin, cannabis, corticosteroids) with no results. At the age of 47 she was diagnosed with a fracture of the right horn of hyoid bone which was surgically removed with no improvement of the symptoms. In 2014 she underwent TC and RMN of head and neck which showed ossification of both stylohyoid ligaments more evident at the right side and a diagnosis of Eagle's syndrome was made. In 2014 the right styloid bone was removed with partial improvement of the symptoms (NRS7). In 2017 the patient came to our attention and a 10KHz spinal cord stimulator was positioned between C2 and C7 with a significant pain relief (NRS4) and a complete relief from foreign body sensation in the throat (Figure 1).

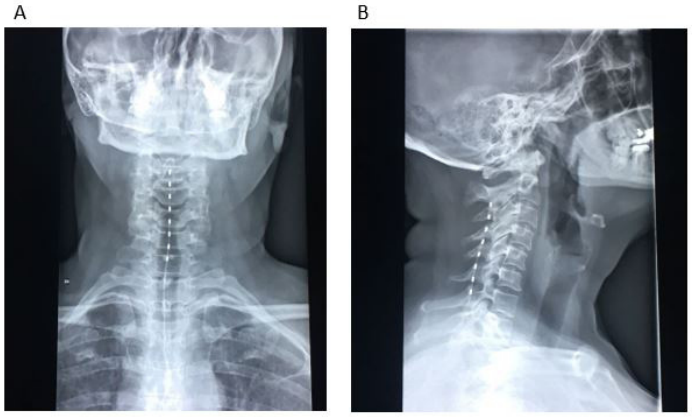

Figure 1: Cervical X-ray image of lead positioning. **A**. Anteroposterior view. **B**. Lateral view. The eight electrodes are spread between C2 and C7.

Discussion

We here reported, to our knowledge, the first case of Eagle's syndrome treated with central neuromodulation performed through Spinal Cord Stimulation (SCS). Our case was refractory not only to the pharmacological treatment but to the surgical approach too. The failure of all therapeutic options could be explained by the long time elapsed between the onset of symptoms and surgical intervention which probably led to a chronic damage of nervous

fibres no more recoverable with the styloid process removal.

Stylohyoid complex is composed of styloid process, stylohyoid ligament and a lesser horn of the hyoid bone. Embryologically, these anatomical structures originate from Reichert's cartilage of the second brachial arch. The styloid process is a pointed part of the temporal bone that serves as an anchor point for several muscles associated with the tongue and larynx.

Eagle considered surgical trauma [tonsillectomy] or local chronic irritation could cause osteitis, periosteitis, or tendonitis of the stylohyoid complex with consequent reactive, ossifying hyperplasia leading to irritation of the structures nearby [1]. Epifanio considered that the ossification of the styloid process was related to endocrine disorders in women at menopause, accompanied by the ossification of ligaments elsewhere [e.g., iliolumbar, thyrohyoid] [16]. Lentini formulated the hypothesis that persistence of the mesenchymal elements [Reichert's cartilage residues] could undergo osseous metaplasia as a consequence of trauma or mechanical stress during the development of the styloid process [17]. Monsour and Young considered that ossification of the stylohyoid ligament complex, caused contraction of the stylopharyngeal muscle and stretching of the XII cranial nerve [18].

A retrospective study by Sekerci et al. indicated that a correlation exists between the presence of a ponticulus posticus [also called an arcuate foramen] and an elongated styloid process. Results were derived using three dimensional (3-D) cone-beam Computed Tomography (CT) scans from 542 patients [19].

Traditional management of Eagle's syndrome includes medical and surgical approaches. Our case showed that SCS should be considered in those cases of Eagle's syndrome refractory to conventional therapies. SCS consists in the placement of electrode leads in the posterior epidural space to electrically stimulate the dorsal columns. There in not yet a certain explanation of its mechanism; the gate control theory has been advocated to explain the effectiveness of SCS [20]. According to the gate control theory, small fibres open the gate, localized at spinal cord level, to let painful stimuli pass to the upper integrating centres of the brain; so nociception, which is the mere process of harmful signals transmission through the nervous system, can become pain i.e., the conscious feeling of an unpleasant sensory and emotional experience. Large fibres, which account for the sensation of touch and vibration, close this gate, thus inhibiting the transit of pain signalling. The electrical stimulation of SCS would preferentially stimulate large fibres, leading to the closure of the gate. This theory doesn't explain why SCS affects mostly neuropathic pain whereas it is less effective on nociceptive pain and why pain relief usually rather lasts even after SCS is turned off [21]. SCS acts even on sympathetic system whose inhibition can improve blood flow; thus, it positively influences pain deriving from peripheral and cardiac ischemia and sympathetic mediated pain as in complex regional pain syndrome (CRPS) [22].

Electrode leads positioning can be surgical or percutaneous; the latter is a less invasive approach and it is performed by the epidural space achievement by a Thuoy needle through which leads are inserted and driven up to the target level, depending on the painful area SCS has to cover: for the posterior occipital region, electrodes are positioned around C2; for upper extremity pain, between C2 to C5; for the hand, C5, C6; for chest wall pain and angina, between T1 to T4; for coverage of thigh and knee pain, the leads are placed between T9 to T10 and for the lower leg and ankle, between T10 to T12. For coverage of the foot, leads are positioned between T11 and L1. Coverage for the sole of the foot can be difficult, and may require stimulation of the L5 or S1 nerve root. The final positions of the leads can be slightly different from these suggested starting points based on patient feedback. The leads are usually placed at the midline or just lateral to the midline.

The electrodes are connected to an implanted pulse generator (IPG) positioned subcutaneously in abdominal region or in the area above the gluteus. Three stimulation parameters can be set: frequency, amplitude, and width. Low frequencies usually range from 40Hz to 125Hz and are associated with paraesthesia; high frequencies [i.e. between 1 KHz and 10 KHz] have the advantage to be paraesthesia free. The width of the electrical field is regulated by the different use of electrodes as cathodes or anodes; the contemporary use of two parallel paramedian leads extends the possibility to shape the electrical field to best cover the painful area; the amplitude correlates with the intensity of stimulation and also affects the extent of electrical field and its depth.

Patients undergoing SCS implant have been generally treated with both pharmacological and non-pharmacological options before they could be considered eligible for such invasive procedure. They experienced a trial stimulation of 2-3 weeks with an external pulse generator in order to assess the effectiveness of SCS before they could be definitively implanted. A pain relief of 50% or greater is considered a reasonable improvement to justify the permanent implant.

Common indications for SCS are Failed Back Surgery Syndrome (FBSS), intractable low back pain, chronic pain of trunk and limbs, chronic reflex sympathetic dystrophy, CRPS, refractory angina pectoris, and peripheral limb ischemia [23-28].

Conclusions

Eagle's syndrome could be sometimes refractory to classical pharmacological and surgical management especially in those patients who received a delayed diagnosis with a consequent chronicization of pain. SCS could be an effective approach in these

challenging situations. This case report opens the way to another promising indication for SCS. This new approach to untreatable Eagle's syndrome has to be validated by other similar experiences and further studies.

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