

Neck-Tongue Syndrome

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Neck-tongue syndrome (NTS) is rare, and characterized by unilateral upper neck or occipital pain and paresthesia in the ipsilateral hemisphere of the tongue due to neck movement. Treatment for NTS is mainly conservative.

Keywords: headache ; manual therapy ; neck pain ; neck-tongue syndrome ; posterior neck pain

1. Introduction

Neck-tongue syndrome (NTS) is a very rare clinical condition defined by the International Headache Society (IHS) as “painful lesions of the cranial nerves and other facial pain” [1]. Characteristic symptoms of NTS include unilateral upper neck or occipital pain, and paresthesia of the ipsilateral half of the tongue due to neck movement [2]. Past studies of prevalence have shown that NTS is a rare case, estimated to occur in 0.22% of adults [3].

According to the reported characteristics of NTS, it can be divided into uncomplicated NTS and complicated NTS [4]. Uncomplicated NTS is related to idiopathic, hereditary, and trauma, and complicated NTS is reported to appear in patients with underlying diseases [5]. For this reason, differential diagnosis is considered very important, and related diseases include cervical artery dysfunction, vertebrobasilar insufficiency (VBI), transient ischemic attack, migraine, ligamentous insufficiency, and Chiari-1 malformation [2][6][7][8].

In the study of Lewis et al. [9], an autosomal dominant genetic pattern was reported through three cases as a genetic trait, and although symptoms were present in adolescence, the symptoms disappeared in adulthood. For this reason, it can be suggested that the pathogenesis according to age may be different. In other words, structural abnormalities in NTS appearing in adults are found, but rarely reported in adolescents. The pathogenesis reported so far is that stimulation of the C2 ventral branch, which is transmitted through the afferent proprioceptive nerve from the lingual nerve to the C2 nerve root and cervical plexus through anastomosis with the sublingual nerve, causes tongue paresthesia [5][10].

Classification of reported treatments for NTS can be divided into pharmacological and non-pharmacological treatments. Pharmacological treatments include antiepileptic drugs, antidepressants, neuropathic pain modulators, and nonsteroidal anti-inflammatory drugs [7][11][12]. Non-pharmacological treatments include cervical collar, physical therapy, exercise, and neuromuscular re-education [4][11][13].

2. Neck-Tongue Syndrome

Table 1 summarizes the characteristic symptoms of the neck and tongue for NTS and the results of the interventions and treatments. In the six case reports [4][13][14][15][16][17], symptoms and treatments were relatively similar. Although patients' ages were similar, most patients were in their 20s or younger, and the number of women was relatively high.

As defined in the study of Lance and Anthony [2], there were unilateral upper neck or occipital pain and paresthesia in the ipsilateral hemisphere of the tongue due to neck movement. This caused neck pain due to the sudden rotation. In addition, accompanying symptoms not reviewed in this study included dysarthria, numbness of the mastoid process region, facial paresthesia, and numbness in the upper extremities [2][7][9]. As a further note, in a systematic review reported in 2018, 39 patients were analyzed [18]. In this study, most of these patients had NTS onset at childhood or adolescence, and those affected were mostly women (56%). Ligament laxity during growth and development and some genetic factors were reported to be causes of the early onset age. In other studies on NTS episodes, there were cases of cervical osteoarthritis [3][7][19], and in magnetic resonance imaging (MRI) studies, there were cases of retrocerebellar arachnoid cysts [20].

Although non-surgical methods for managing NTS are common, a study on the surgical management of NTS confirmed that symptoms were relieved by resolving the compression on the C2 spinal nerve by the protuberant atlantoaxial joint [21].

In the pharmacological and non-pharmacological interventions included in non-surgical interventions, pharmacological interventions were performed only in the Allen et al. [17] study. In the other five case reports, manual therapy, exercise, and cervical collar were found to be effective.

Differential diagnoses of idiopathic and post-traumatic NTS can be made by considering the accompanying symptoms. Treatment may include using a neck collar or medication to relieve symptoms, and manual therapy and exercise appear to be appropriate as an intervention. However, in a review by Gelfand et al. [18], pathologically, when ankylosing spondylitis and rheumatoid arthritis are accompanied by chronic pain and joint instability in the cervical spine (C1, C2), it is difficult to explain the pain of NTS neurologically. Furthermore, what can be said to be different from our study is that there is no advantage of spinal manipulation therapy for the type of suspected instability. However, in childhood and adolescence cases, there are cases of compression due to shortness of the scalenus and congenital abnormalities or shortness of the sternocleidomastoid muscle. At this time, symptoms are improved through the effect of manual therapy for the absence of neuromuscular control development of muscles and ligaments due to neurophysiological pathology and structural instability [22]. Therefore, it is suggested that interventions such as spinal manipulation therapy, manual therapy, exercise, and neuromuscular re-education can be helpful as a conservative treatment method.

Although it is a syndrome with a relatively low incidence and long-term discomfort, randomized controlled trials are difficult to conduct, but it is worth studying the most effective management through a long-term cohort study. In addition, in items b, c, and d of ICHD-3, first, diagnosis of intermittent pain rather than chronic pain when there is no neck movement, second, sharp pain during rotation, accompanied by paresthesia of the tongue, and third, there is partial agreement as it is essential for the diagnosis of NTS pain that the duration of treatment ranges from a few seconds to a few minutes.

Table 1. Characteristics of the case reports.

Study	Sex	Age	Symptoms			Interventions			Results	Author's Conclusion
			Triggers	Neck	Tongue	NTS Duration	Types	Intensity		
Roberts. [14]	Female	34	Rotation of the head to the left side	Mild pain in the left suboccipital region	Described as "tingling," "shooting," "piercing," and "electric shock-like" on the left side tongue	>2 years	Spinal manipulative therapy, myofascial release, and exercises	Two weeks	Symptoms improved after 2 weeks of intervention and persisted after 2 years	Chiropractic care is helpful for NTS
Niethamer and Myers. [13]	Female	13	Turning head to one side or the other or looking up	Sharp pain	Numbness/tingling of the ipsilateral tongue	> 6 months	Manual therapy, exercise (including postural awareness)	Eight visits	Patient-specific functional scale, cervical ROM, postural normalization	An effective known treatment for cervicogenic headache may be the basis for treating NTS
Borody. [15]	Female	24	Brisk active rotation	Upper neck (right) "sharp" or "piercing" pain	Ipsilateral tongue numbness	Recurrent pain after 8 years of age	Diversified spinal manipulative therapy performed to the restrictions throughout the cervical spine	Various intervals	Reduction in the frequency and intensity of the symptoms	In the absence of other additional symptoms and upper cervical instability, spinal manipulative therapy is effective
Chirchiglia et al. [4]	Female	42	Rotation of the head to the right side	Severe neck pain	Paresthesia of the right tongue	A month later, reported a whiplash injury, following a car accident	Topiramate combined with non-steroidal anti-inflammatory drugs and cervical collar	Two weeks	Improved pain both in the neck and the tongue, reducing the episodes in frequency and intensity	Pharmacological treatment is prophylactic

Study	Sex	Age	Symptoms			Interventions			Results	Author's Conclusion
			Triggers	Neck	Tongue	NTS Duration	Types	Intensity		
References										
Chu and Lin, [16]	Male	47	Head-turning to the right, forward neck pain or chewing	right suboccipital pain	Tingling of the right face and ipsilateral hemihand	>3 months	Chiropractic regimen	Three times a week for four weeks	The painful episodes had disappeared and the active range of neck motion had increased	Conservative management, including physiotherapy and minor cervical adjustment, may be the preferred initial treatment
1. Hu, N.; Dougherty, C.										
Neck-tongue syndrome. Curr. Pain Headache Rep. 2016, 20, 27.										
2. Lance, J.W.; Anthony, M.										
Neck-tongue syndrome on sudden turning of the head. J. Neurol. Neurosurg. Psychiatr. 1980, 43, 97–101.										
3. Sjaastad, C.; Bakkevig, M.	Female	11	Head-turning	Severe shooting/burning pain in the posterior neck	Ipsilateral hemihand numbness	>5 months	Gabapentin and topical lidocaine		Less pain intensity at follow-up after 1 year	It is characterized by additional symptoms of headache
4. Chirchiglia, D.; Della Torre, A.; Chirchiglia, P.; Pugliese, D.; Gabriele, D.			Sudden whiplash injury							
Interdisorder Neurosurg. 2019, 15, 38–40.										
5. Bogduk, N.	Female	11	Sudden turning/neck movement/fatigue	Pain in the posterior neck	Ipsilateral numbness	After symptom onset		4 years follow-up	Relieve symptoms naturally	headache was spontaneously resolved in 2 patients and
6. Evans, R.W.			Sudden onset of head-turning	Severe burning pain in the posterior neck	Ipsilateral tightness/twisting	Some months		6 years follow-up		patient was relieved of headaches by gabapentin
7. Fortin, C.J.; Biller, J.	Male	12								
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