The neck-tongue syndrome

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Abstract
The neck-tongue syndrome, consisting of pain in the neck and altered sensation in the ipsilateral half of the tongue aggra-vated by neck movement, has been attributed to damage to lingual afferent fibres travelling in the hypoglossal nerve to the C2 spinal roots. The lingual afferents in the hypoglossal nerve are thought to be proprioceptive. Two further cases of the neck-tongue syndrome are described, the spectrum of its clinical manifestations is explored, and the phenomenon of lingual pseudoanesthesia is illustrated as a result of the presumed lingual deafferentation.

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Lance and Anthony1 described four patients, aged 8 to 14 years at onset, with a brief (seconds to a minute) sharp pain on one side of the upper neck or occiput on sudden rotation of the neck, followed immediately by transient (seconds to minutes) ipsilateral numbness of the tongue. Two of these cases had congenital anomalies of the upper cervical spine on radiography. Cyriax2 had previously noted a similar syndrome in two patients, one of whom was a 45-year-old woman with attacks of severe pain at the occiput and left side of the neck and numbness of the tongue and back of the palate, precipitated by turning her head sharply to the right. The other patient had a cervical disc lesion and a sensation of pins and needles felt in the tongue only.

Lance and Anthony1 concluded that the pain was due to irritation of the second (and third) cervical root, which is vulnerable to compression in its course between the atlas and axis during neck rotation. They explained the unilateral numbness of the tongue on the basis of afferent impulses (notably, proprioceptive) travelling from the lingual nerve via the hypoglossal nerve to the second cervical root.

Bogduk3 clarified the anatomical basis for the syndrome by cadaveric dissection, demonstrating that the C2 ganglion and spinal nerve lie dorsal to the lateral atlantoaxial joint; the joint is innervated by the C2 ventral ramus. He proposed that the pain was due to temporary abnormal subluxation of the lateral atlantoaxial joint on rotation of the head. This strains the joint capsule, and thereby causes pain in the suboccipital region. He attributed the unilateral numbness of the tongue (and of the skin behind the ear) to compression of the C2 ventral ramus by movement or subluxation of the lateral atlantoaxial joint. He suggested that the abnormal sensation on compression of proprioceptive afferents from the tongue is similar to the altered sensation or numbness recognised in Bell’s palsy, despite intact trigeminal sensation, due to compression of proprioceptive fibres in the facial nerve.

Since these original descriptions of the neck-tongue syndrome, a few other cases have been described. Lance4 briefly mentions a further four patients in whom the syndrome started in late childhood or adolescence. Elisevich et al5 reported a 53-year-old woman with assimilation of the atlas to the occiput presenting with neck pain and ipsilateral paraesthesiae of the tongue on head turning, in whom the symptoms became persistent and incapacitating. At operation, the C2 spinal nerves were found to be compressed by a protuberant atlantoaxial joint. Resection of the superficial parts of the C2 spinal nerves led to partial relief.

Webb et al6 described three cases with ipsilateral neck pain and tongue numbness precipitated by neck movement, first developing symptoms at ages of 10 to 61 years. Degenerative changes of the anterior atlantoaxial joint were present in two of the cases. Restriction of neck movement, including use of a surgical collar in two of the cases, controlled the symptoms.

Fortin and Biller7 described a 57-year-old woman with repeated episodes of numbness of the posterior right half of the tongue and right posterolateral oropharynx, an ipsilateral “ear pressure”, and a sharp aching pain radiating along the border of the trapezius to the acromioclavicular joint. At the same time she noticed a pressure sensation in the occiput and paraesthesiae in the fourth and fifth fingers of the right hand. The episodes lasted one minute, and she herself noted no clear precipitant, but they were precipitated by extreme lateral rotation of the neck. The region of the atlantoaxial joint was normal radiologically. They also described a 29-year-old man, with previous head injury and lower thoracic vertebral compression fractures after a motorcycle accident, who complained of mainly right, but sometimes left sided (never both sides simultaneously), hemiglossal and facial paraesthesiae, sharp occipital pain shooting to the periorbital area, dysarthria, nasal...
congestion, sialorrhoea, and paraesthesiae in the ulnar distribution of the upper limb. The symptoms in both patients were relieved by use of a surgical collar.

Bertoft and Westerberg\(^5\) reported a further six patients with a complete syndrome, and three with an incomplete syndrome, which they define as having characteristic attacks, but lacking the paroxysmal numbness of the tongue. We have not included these cases of an incomplete syndrome in the present discussion, but two of the patients with a complete syndrome had radiological evidence of arthritis of the lateral atlantoaxial joints, and in one the symptoms were abolished after surgical atlantoaxial fusion.

Cassidy et al.\(^6\) described three patients with the neck-tongue syndrome, who were successfully treated with rotational manipulation of the cervical spine. Terrett et al.\(^7\) described eight cases of the neck-tongue syndrome, which were treated with spinal manipulative therapy. They also referred to an additional 54-year-old woman with headaches, who, after cervical manipulation, developed left-sided tongue numbness, persisting for two years, together with an intermittent dysarthria. Hankey\(^8\) described a more classical neck-tongue syndrome in a 14-year-old girl with recurrent episodes of unilateral upper nuchal pain radiating to the ipsilateral side of the tongue on ipsilateral rotation of the neck.

Noda and Umezaki\(^9\) described a 35-year-old woman with paroxysmal episodes, lasting up to one minute, of numbness of the left neck and occiput, with simultaneous numbness of the left half of the tongue, preceded by tonic spasm of the left arm. They suggest this is a component of a spinal tonic seizure related to an upper cervical inflammatory myelopathy involving the dorsal funiculus, proximal to the C2 and C3 dorsal root ganglia, and hence a spinal neck-tongue syndrome.

In total, 36 cases of this syndrome have been previously described in the literature, some very briefly,\(^10\) although Bogduk\(^11\) comments that on discussion with other neurologists and neurosurgeons it seems that the syndrome is not rare. Accordingly we describe two further cases of the syndrome, due in one case to atlantoaxial osteoarthritis, and in the other to a traumatic fracture and displacement of the occipital condyle. A striking feature was dysarthria and, in the second case, pseudoathetosis of the tongue.

### Patients

#### CASE 1

A 51-year-old woman complained of a progressive stiffness and pain in the neck for six years. Three years previously she had noticed that the pain was exacerbated by movement of the neck to a critical position, with the chin down and to the right. She was aware of a grating sensation in the neck, and the pain appeared to be deep to the angle of the jaw on the left, radiating to the occiput. These exacerbations were associated with a tingling sensation of the left side of the tongue, and a sensation of the tongue being pulled into the back of the throat on the left, causing a feeling of choking, with difficulty speaking.

Neurological examination was normal, and in particular touch and pin prick sensation on both sides of the tongue was preserved. The tongue and palate appeared normal, with normal movement. Cervical spine movement was severely limited due to pain.

A cervical spine radiograph showed degenerative changes in the mid and lower cervical spine, with rotation at the atlantoaxial joint, but no horizontal subluxation. A cervical spine CT scan with the head turned to the right and left showed no structural lesion or abnormal movement. Rotatory movement was equal to the right and left, with no dislocation of the lateral atlantoaxial joint. The rotation was about 40 degrees on each side, a little more than usually seen, but probably not pathological. A radioisotope bone scan, however, revealed increased uptake in the left atlantoaxial joint, attributed to osteoarthritis. Injection of a local anaesthetic and steroid into this joint led to brief relief of the pain.

#### CASE 2

A 32-year-old woman accidentally fell down a flight of 12 stairs at night, hitting the right frontoparietal region of her head against a table. This caused a scalp laceration but no loss of consciousness or immediately apparent neurological deficit. The next day she awoke with a continuous right sided occipital headache and neck pain exacerbated by attempts at neck movement. She had slurring of speech, continuous altered sensation of the tongue and pharynx on the right with a feeling of the tongue moving to the left, and difficulty manipulating food in the mouth.

On examination she had tenderness over the upper cervical spine, and neck movements were severely restricted by pain. The tongue was rotated anticlockwise in the mouth, with consequent appearance of decreased bulk of the left side of the tongue (fig A). On forward protrusion of the tongue there was equal bulk of the two sides, but great difficulty in maintaining a sustained steady forward position, the tongue moving involuntarily to right and left (fig B). There were no visible fasciculations. There was full movement of the tongue to the left (fig C) despite partial denervation of the right half of the tongue demonstrated by EMG. Tongue protrusion to the right was incomplete and poorly sustained although no weakness of the left half of the tongue could be detected (fig D). On fibreoptic endoscopy the base of the tongue was more posterior than usual, and partially obstructing the larynx. She had a mild lingual dysarthria. Sensation of the tongue and pharynx to touch, taste, and pin prick was normal. Neurological examination was otherwise normal.

Cervical spine radiography was normal. An upper cervical CT scan was normal, but a CT scan of the skull base showed a fracture of the
right occipital condyle, with slight medial displacement of the fragment just distal to the right hypoglossal canal.

Discussion
The clinical spectrum of the neck-tongue syndrome, including age at presentation, chronicity, and disablement, has broadened since the original description. The two patients we describe had a feeling of abnormal posturing or movement of the tongue as a significant feature of their symptomatology.

In case 2 the positioning of the tongue was clearly abnormal and she was unable to maintain a sustained normal posture of the tongue in the absence of any specific weakness. In particular there was no weakness of tongue movement to the left, which would be affected by motor denervation of the right side of the tongue. We propose that this is a result of proprioceptive deafferentation of the tongue, with the clinical sign of lingual pseudoathetosis. This is similar to pseudoathetosis as previously described in the limbs.14 Pseudoathetoid movements of the tongue seem particularly complex in the absence of any restriction of direction of movement imposed by a joint. The associated dysarthria has features of both impaired lingual control and pharyngeal obstruction as manifestations of lingual pseudoathetosis predominantly of the tip and base respectively.

Proprioception in the tongue has been studied in animals and humans,19 with controversy initially concerning whether the tongue musculature has proprioception. As a generalisation, it seems that muscle spindles are not found in the tongue of mammals, except in moles, primates, and humans.16 Muscle spindles have been demonstrated histologically in the human tongue.17,18 The afferent pathways seem to be in the hypoglossal nerve,19,20 which is otherwise thought of as being a purely motor nerve.

Lingual pseudoathetosis is suggested in previous descriptions of neck-tongue syndrome. A 15-year-old boy had a right occipital pain and numbness of the right side of the tongue on sudden neck rotation, and a feeling that the tongue was twisted sideways in his mouth.1 The symptoms lasted a few seconds only. A 29-year-old man had ipsilateral occipital pain and tingling in his tongue, with dysarthria and nasal congestion, precipitated by neck movement.7 A 60-year-old man, with symptoms since the age of 10 years, had experienced a sensation of his tongue being twisted sideways in the mouth during an attack, “as though the whole tongue is cleaving to the roof of the mouth”.4 A 19-year-old woman with pain in the left side of the neck on sudden head turning also noticed deviation of her tongue to the right for 10 seconds, with anaesthesia.4 A 53-year-old woman described paralysis of her tongue for about five seconds on sharp rotation of the neck.10 A 19-year-old woman described her tongue as seeming to take up all the room in her mouth, having a mind of its own, being uncontrollable, and resulting in dysarthria.10 A 46-year-old man experienced the symptoms of the neck-tongue syndrome, his tongue being numb and rigid, with dysarthria.10 A 26-year-old woman experienced the neck-tongue syndrome, with an inability to move her tongue.17 In these cases the dysarthria, as in case 2, may be related to proprioceptive deafferentation rather than a primary motor deficit.

A 53-year-old woman had pain in the right side of her neck and tingling of the ipsilateral side of her tongue, initially intermittent but then constant. This was aggravated by head turning and talking, and when severe the pain radiated to the back of her head or right arm and caused mild dysphagia related to a feeling of muscle spasm in the throat.1 A 34-year-old man complained of intermittent left sided neck pain radiating to the occiput, with ipsilateral aching and numbers of the tongue. The symptoms were precipitated by rapid
neck flexion and rotation to either side when
breathing during competitive free-style swim-
mimg. Occasionally the symptoms were so
intense and associated with a choking sensa-
tion that he had to stop swimming during a
race. (He had experienced the same symp-
toms while swimming between the ages of 10
and 22 years).  

Most reported cases have had transient
symptoms and signs, but in case 2 these 
were constant, enabling the tongue posture and
movement to be observed. The tongue
seemed to be drawn back in the mouth, com-
patible with the symptom described in case 1,
and possibly explaining the muscle spasm in
the throat of the 53-year-old woman6 and 34-
year-old swimmer.  

Of the cases of the neck-tongue syndrome
previously described, four involved abnormal-
ities at the atlantoaxial joints, two fusion of
the atlas to the occiput, and another a minor
abnormality of the occipital condylar
processes. A cervical disc lesion was noted in
another. In the other cases no clear pathology
of the upper cervical joints was noted
(although in one an intrinsic myelopathy was
suggested). In case 1 the structural abnormal-
ity was osteoarthritis at the left atlantoaxial
joint demonstrated by radioisotope bone
scan. Irregular loss of the left lateral
atlantoaxial joint space with reactive sclerosis
and osteophytic lipping was noted in a 36-
year-old man,4 and moderate degenerative
changes at the anterior atlantoaxial joint in a
65-year-old woman.5  

Another possible example of the condition
was described by Lees et al20 as “paroxysmal
hemiglossal twisting”. These authors reported
two patients, and tentatively suggested dysto-
nia or tonic seizure as the cause. The first
case was a 61-year-old woman who, on
review of the original records, described a tin-
gling sensation at the back of the tongue on
the left, the tongue then deviated to the right,
and she was unable to move her tongue to the
left, associated with dysarthria. On other
occasions the tongue twisted from side to
side, and she had bitten the left side of her
tongue several times. Touch sensation of the
tongue was normal during an episode. She
had repeated episodes, lasting for about one
minute, which were infrequent, with no clear
precipitating neck movement, but appeared
to be related to exercise. The attacks could be
associated with discomfort down the left side
of the neck and behind the left ear. A cervical
spine radiograph showed no abnormality of
the upper cervical region, and CT scan of the
atlantoaxial joint was normal. Except for the
absence of precipitation by neck movement,
this case has features in common with the
neck-tongue syndrome, and the abnormal
twisting of the tongue may be a lingual
pseudoarthetosis due to lingual proprioceptive
deafferentation, with associated C2 pain.
(The second patient had brief spontaneous
attacks of tongue twisting and dysarthria
without pain or paraesthesiae).  

In case 1, the site of deafferentation is
assumed to be in the neck, related to the
atlantoaxial joint. In case 2, the site of deaf-
ferentation is uncertain, and could be at the
atlantoaxial joint related to the fall and twist-
ing of the neck, related to the displaced
occipital condyle and abnormal atlanto-
occipital joint, or in the hypoglossal nerve
after exit from the hypoglossal canal related
to the displaced bone fragment. Previous
abnormalities of the atlanto-occipital joint
were described in a 26-year-old man with
anteromedial bosses of bone on the condylar
processes of the occiput representing a minor
anomaly of the occipital assimilation.7 The
spinous processes of the atlas were fused to
the adjoining occipital bones in a 15-year-old
girl,1 and assimilation of the atlas to the
occiput was noted in a 53-year-old woman.5  

Thus of the 38 cases now described,
pathology of the occipitoatlantoaxial joints
has been noted in 10.

The onset of symptoms in case 2 was
clearly related to trauma, as were 10 previous
cases. A 29-year-old man had a motorcycle
accident, with head and thoracic spine injury,
two years before developing the neck-tongue
syndrome.7 A 26-year-old man was hit in the
midfrontal region by a child’s swing, with lac-
eration of the forehead. Shortly afterwards
he developed numbness and weakness of the
right hand lasting for several days. Ever since
he has had transient symptoms of numbness
of the right side of the tongue and right
fourth and fifth fingers, and right sided neck
pain, lasting a few seconds only, on sudden
rotation of the neck.  

A 36-year-old man acciden-
tially jarred his neck, and subsequently,
whenever jolted unexpectedly, experienced
high cervical pain radiating to the left side of
his tongue “like the aftermath of a dental
anaesthetic”.6 A 28-year-old woman devel-
oped the neck-tongue syndrome the day after
a whiplash injury, which persisted for one
year, but resolved with spinal manipulation.4
Other cases associated with trauma include
four after a motor vehicle collision, and one
developing eight years after a water skiing
accident.8  

The transient relief of the symptoms in
case 1 by injection of local anaesthetic and
steroid into the lateral atlantoaxial joint is
supportive of the symptoms being related to
this joint, and raises the possibility of treat-
ment by permanent cervical fusion. Previous-
ly reported surgery with bilateral resection of
C2 spinal nerves with their dorsal and ventral
rami, initially relieved the symp-
toms, but after one week there was a recur-
rence of mild tingling sensations in the
tongue when the patient was tired. The neck
pain also recurred, but less severely than
before the operation. The authors suggested
that the recurrence of tongue symptoms
could be explained by facilitation of residual
proprioceptive fibres in the adjacent unin-
jured spinal roots. In another patient, surgical
atlantoaxial fusion abolished the symptoms.9
Other patients have had their symptoms
relieved by spinal manipulation.9,10 Symptoms
have been improved by preventing the precip-
itative neck movement with a surgical collar,
but the benefit is not necessarily permanent, and with no long term follow up of this syndrome the natural history is unclear.

Since the initial description by Lance and Anthony1 of patients aged 15 to 26 years, with an onset age of eight to 15 years, it has become apparent that the same syndrome may present up to at least the age of 61 years, and may present as transient symptoms which may persist, may resolve and relapse, or may become permanent and present as a permanent deficit. In a significant proportion there is pathology at the atlantoaxial and atlanto-occipital joints, and in some cases the syndrome may result from trauma.

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