

An Unusual Cause of the Neck-Tongue Syndrome

To the Editor:

The neck-tongue syndrome (NTS) is a rare clinical entity characterized by paroxysmic episodes of intense pain in upper cervical or occipital area associated with disorders of ipsilateral hemi-tongue presenting as numbness, pseudoathetosis, dysarthria, and lingual paralysis aggravated with neck movement^{1,2}. We describe a case of NTS with an unusual etiology.

A 74-year-old woman presented with severe pain at the back of the neck that improved spontaneously within 2 weeks. During this time she developed dysarthria and was observed to have a left-sided lingual paralysis. Systemic enquiry was unremarkable. No other focal neurological abnormality was detected. She had marked nodal osteoarthritis in her hands and her cervical spine rotations were reduced. There was no clinical evidence of inflammatory arthritis. Her dysarthria gradually improved but never resolved completely.

Blood tests revealed a normal inflammatory, biochemical, and immunological screen. Magnetic resonance imaging and computed tomography (CT) scan of the cervical spine showed degenerative changes with predominant involvement of the left atlantoaxial joint and odontoid process. Prominent osteophytes, juxtaarticular sclerosis, and marked erosive arthropathic changes were noted at this level (Figure 1A). The left side of the tongue was atrophic with fatty infiltration, in keeping with changes of left hypoglossal nerve denervation (Figure 1B). An intracranial pathology was ruled out.

The hypoglossal nerve, after exiting the hypoglossal foramen, courses close to the atlantoaxial joint³. We postulate that in our patient, the extensive erosive arthropathy and osteophytosis impinged on the left hypoglossal nerve, explaining her symptoms. Erosive arthropathy was not limited to the atlantoaxial joint; erosive facet arthropathy was also seen at several levels throughout the cervical spine. There was no clinical or serologic evidence of rheumatoid arthritis. Hence, the most plausible diagnosis in our patient with an erosive arthropathy would be erosive osteoarthritis or crystal arthropathy associated with nodal osteoarthritis. Between these 2 plausible causes, crystal arthropathy secondary to calcium pyrophosphate deposition disease (CPPD) has been described in the literature as causing extensive erosive changes, bony sclerosis, osteophyte formation, and a pseudo-Charcot type appearance in the cervical spine, particularly at the craniocervical junction, very similar to the findings in our case⁴. Hydroxyapatite deposition disease, gout, and erosive osteoarthritis usually do not present with such extensive radiological changes involving the cervical spine.

There are 2 categories of NTS: complicated NTS due to the presence of an underlying disease process (inflammatory or degenerative) and uncomplicated NTS (idiopathic or trauma-related)⁵. The originally proposed anatomical explanation for the syndrome, interconnections between the lingual and hypoglossal nerves and the C2, C3 nerve roots, is consistent with this syndrome complex arising from arthritis affecting the C1-C2-C3 artic-

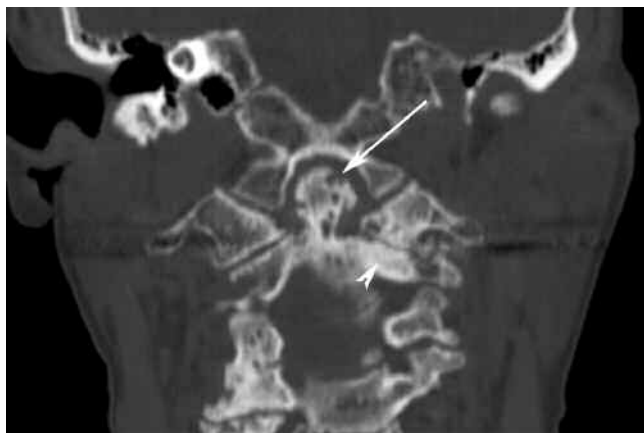


Figure 1A. Coronal reformatted CT image showing sclerosis and narrowing of the left atlantoaxial joint (arrowhead) and erosions of the odontoid process (arrow).

ulation⁶. The lingual pseudoathetosis is assumed to be due to a proprioceptive deafferentation of the tongue⁷. Etiologies causing NTS in adults reported in the literature include congenital anomalies of cervical spine, degenerative spine disease, ankylosing spondylitis, tuberculous atlantoaxial osteoarthritis, and head or neck trauma. To our knowledge, complicated NTS consequent to erosive CPPD arthropathy has not been described before, and readers should be aware of such a possible link.

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Figure 1B. Coronal T1-weighted image showing fatty change in the left hemi-tongue as evidenced by increased signal in that area (arrow).