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IDIOPATHIC UNILATERAL PERSISTENT HYPOGLOSSAL NERVE PALSY

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ABSTRACT

When combined with other cranial nerve palsies or additional illness, hypoglossal nerve palsy is a common finding in neurological disorders, and it has distinct clinical symptoms, such as unilateral atrophy of the tongue muscle. It might be the first or only symptom of an intracranial or extracranial space-occupying lesion, a head/neck injury, or an internal carotid artery vascular anomaly. Idiopathic isolated unilateral hypoglossal nerve palsy (HNP) is a rare occurrence that should be detected by exclusion. To our knowledge, this is the first comprehensive assessment of the literature on idiopathic isolated unilateral HNP. This is a fairly unusual illness, but it should be considered for diagnosis. It necessitates a methodical and step-by-step approach to etiological diagnosis. We evaluate documented examples in the literature in this paper, emphasizing the importance of a segmental and sequential approach in the diagnosis and treatment of this illness.

Database: Google scholar, PubMed.

KEYWORDS: hypoglossal nerve palsy, idiopathic, unilateral.

INTRODUCTION

The hypoglossal nerve (cranial nerve XII) is a purely motor nerve that innervates the tongue's intrinsic and extrinsic muscles, as well as the anterior neck's strap muscles. Classic clinical signs of hypoglossal nerve palsy include tongue deviation to the afflicted side, atrophy, and fasciculation of the tongue muscle. Hypoglossal nerve palsy is an uncommon ailment with little documentation in the literature.[1]

The nucleus and intramedullary filaments, cisternal area, hypoglossal channel (skull base), nasopharyngeal/ oropharyngeal carotid space, and sublingual area are the five areas of the hypoglossal nerve. A distinct illness generally affects each location.[2] A tumour is the most prevalent cause, followed by trauma, stroke, and infection, with 3% of cases being described as idiopathic. Only a few cases of isolated HNP have been documented following influenza vaccination or a common cold, as well as a few cases of infectious mononucleosis. HNP usually involves other cranial nerves, and isolated HNP that does not include other cranial nerves or long tracts is uncommon.[3] Idiopathic unilateral HNP is extremely uncommon, however numerous examples of transitory or chronic HNP have been described. According to some researchers, Self-limiting idiopathic HNP is thought to be identical to Bell's palsy of the facial nerve.[4]

Anatomy and Aetiology

The 12th cranial nerve, the hypoglossal nerve, innervates all tongue muscles save the palatoglossus. It exits the hypoglossal canal after leaving the medulla and passing along the base of the skull. It goes inferiorly deep to the internal carotid artery and internal jugular vein after entering the nasopharyngeal region. Before entering the tongue, the nerve passes beneath the digastric muscle.[5] The hypoglossal nerve's route can be split into segments, each of which is vulnerable to distinct sorts of disease, leading in nerve palsy. The sublingual and carotid spaces, the skull base, the cisternal space, and the medullary space are the segments in question.[6] Tumors (49 percent), trauma (12 percent), cerebrovascular accident (6 percent), infection (5 percent), and multiple sclerosis (5 percent) are the most prevalent causes of isolated hypoglossal nerve palsy.[5] Only 3% of instances have been proven to be idiopathic, and only four cases of unresolving idiopathic hypoglossal nerve palsy have been recorded in the literature.[7]

The hypoglossal nuclei placed in a paramedian position inside the medulla oblongata correspond to the first or medullary segment, generating a protrusion into the fourth ventricle known as the hypoglossal eminence.[8] The nerve fibres escape the medulla in the pre-olivary sulcus and extend anteriorly, lateral to the medial lemniscus. The nerve exits as a series of rootlets that converge to create the hypoglossal nerve.[9] The hypoglossal nerve's second or cisternal segment runs

along the pre-medullary cistern, anterior to the posterior inferior cerebellar artery and posterolateral to the vertebral artery.[10] The nerve enters the hypoglossal canal from the cisternal segment, which is positioned between the occipital condyle inferiorly and the jugular tubercle superolaterally. The hypoglossal nerve's third segment, or skull base segment, is located here.[11]

The cranial nerve XII is positioned medial to the glossopharyngeal, vagus, and auxiliary cranial nerves in its fourth or nasopharyngeal carotid segment.[10] The nerve loops anteriorly, inferior to the posterior belly of the digastric muscle, at the level of the mandible's angle, and then anteriorly along the surface of the hyoglossus muscle at the level of the hyoid bone. This relates to the fifth or sublingual segment, which lies along the surface of the genioglossus muscle before piercing it.[12]

The hypoglossal nerve has a meningeal branch as well as anastomotic branches with the superior cervical sympathetic ganglion, the cervical plexus, and the inferior ganglion of the vagus nerve. It also has branches that anastomose with C1–C3 nerves.[11] A collection of fibres from C1 that runs down the carotidspace with the hypoglossal nerve form the ansa hypoglossi. It provides motor innervation to the omohyoid, sternothyroid, and sternohyoid muscles of the infrahyoid strap.[13] There are other small branches that connect to the 5th cranial nerve's V3 branch and the lingual nerve. Within the hypoglossal canal, the basilar venous plexus surrounds the hypoglossal nerve. It generally thickens and spreads into the nasopharyngeal cavity.[14] The tongue deviates contralateral to the side of the lesion when the disease affects the supranuclear level of the 12th cranial nerve. The symptoms are ipsilateral if the disease affects the nerve's nuclear or infranuclear levels. The tongue deviates to the side of the lesion, which is accompanied by unilateral atrophy and fasciculations.[15] Chronic hypoglossal denervation results in muscle volume loss, fatty replacement, hemiatrophy on the weak side, and tongue prolapse into the oropharynx.[16] Unilateral alterations of the tongue musculature with low signal intensity on T1, high signal intensity on T2, and aberrant enhancement were seen on MRI. It appears with edoema, a strong signal on both sequences that correlates to fatty infiltration, and modest enhancement during subacute periods.[17]

Diagnosis

Idiopathic hypoglossal palsy is diagnosed by ruling out all other possible causes. Other causes of the palsy must be ruled out before such a diagnosis can be established.[18] To rule out curable illnesses, meticulous step-by-step examinations should be carried out. Radiological and haematological tests are among them. MR scans of the head and neck, as well as a chest radiograph, are used in radiological studies. As part of routine screening and to evaluate infective reasons, haematological studies include complete blood count, urea and electrolytes, C reactive protein, erythrocyte sedimentation rate, and viral serology. autoimmune antibodies and general immunology to rule out an immunological cause; blood glucose to rule out diabetes; haematinics to rule out dietary shortages; autoimmune antibodies and general immunology to rule out an immunological cause.[5]

Review of literature

From personal experience over a 26-year period, Keane et al.[19] recorded 100 occurrences of hypoglossal nerve palsy. Only three of them were idiopathic, and they all healed on their own. Patients varied in age from 8 to 73 years old. Nine instances of solitary hypoglossal nerve palsy were documented by Combarros et al.[20], five of which had identifiable aetiology and four of which were idiopathic. By five months after presentation, all idiopathic patients had entirely recovered. According to Sayana et al.[6] A 76-year-old Caucasian lady was referred with a three-week history of dysphagia and speech impairment. There was no previous history of trauma, surgery, infection, or a CVA. All of the studies were uneventful, and there was no evidence of a causal cause. The palsy in our case was chronic and lasted four years. Ho et al.[7] described a case of a 25-year-old Caucasian woman who appeared with a right-sided tongue deviation that had started five years prior to her presentation. She had struggled with speaking and reading aloud in her previous job as a schoolteacher. A wide variety of examinations yielded ordinary results once again. Despite the fact that the palsy remained unresolved, this patient was able to modify her speech and mastication with time, thus the palsy had no long-term functional consequences.

Bagan-Sebastian et al.[21] described a 24-year-old lady who had a 10-year history of tongue motility problems and dysarthria. There was no evident cause, and this patient adjusted well to the scenario, with only minor long-term functioning issues. Freedman et al.[5] described a 22-year-old male patient who had been having trouble moving and controlling his tongue for 18 months. He was in good physical condition. His hypoglossal nerve palsy has no identifiable cause.

DISCUSSION

Dysarthria, drooling, and dysphagia are the most common symptoms.[22] On clinical examination, tongue protrusion to the afflicted side, muscle atrophy, and varied fasciculation are all present. The majority of hypoglossal nerve palsies go away on their own. Cases that last for a long time are exceedingly rare. The symptoms in chronic instances do not deteriorate over time, according to the literature. These instances have been treated conservatively, with referrals to speech and language therapy. It's important to note that if the symptoms increase over time, a repeat and comprehensive clinical examination, as well as the relevant tests, are required to confirm the diagnosis.

The 12th cranial nerve, the hypoglossal nerve, supplies motor innervation to the tongue's intrinsic and extrinsic muscles. The motor component of the hypoglossal nerve, on the other hand, is extremely complicated and little known.[5] It leaves the skull and enters the nasopharyngeal cavity, where it travels inferiorly and profoundly to the internal carotid artery and internal jugular vein, before passing beneath the digastric muscle and into the tongue's substance.[23] Because this neuropathy is often associated with other neurological disorders, pure HNP is uncommon. Vascular illnesses, such as median medullary syndrome and carotid artery dissection, are the most common causes of hypoglossal nucleus dysfunction.[24] Primary and metastatic tumours are the most prevalent causes of hypoglossal canal obstruction, and HNP is commonly linked with vagus nerve and accessory nerve dysfunction. Furthermore, isolated HNP may be the first or only indicator of a more serious underlying condition. As a result, practitioners should be wary in such situations and be prepared to take an investigative approach to determine the underlying condition.[5] Our HNP instances were provided without any historical context. Few comprehensive evaluations of HNP cases exist, including bilateral HNP and instances including additional cranial nerve palsies. A retrospective investigation of 100 instances of HNP was described by Keane.[19] According to the author, the most prevalent cause, was a tumour (almost 50% of cases), followed by trauma (12%), stroke (6%), and infection (4%), with 3% of cases being classified as idiopathic. Stino[23] published a retrospective investigation of 245 instances with HNP, revealing the following etiological categories: postoperative (29.3% of cases), malignancies (27.2%), inflammatory (7.3%), radiation-induced (6.1%), traumatic (4.1%), and idiopathic (15.1 percent). In a French paper, Tommasi-Davenas et al. reported on a retrospective investigation of 32 instances with HNP. According to the survey, the most prevalent cause was a tumour (53.1%), followed by vascular difficulties (18.9%), trauma (12.5%), and inflammation (12.5%). Isolated HNP is caused by primary or secondary malignancies of the skull base, vascular diseases, iatrogenic causes, and inflammation in combination, and idiopathic isolated unilateral HNP is uncommon. Regrettably, these studies did not include information from specific situations.

Ipsilateral headache (33.3%), fever with or without common cold (16.7%), and a neck muscular pull(4.2 percent) were the most prevalent prodromal symptoms of idiopathic isolated unilateral HNP. It's worth noting that half of the patients didn't have any prodromal symptoms. Idiopathic isolated unilateral HNP cases reported fewer headaches (33.3%) than entire HNP cases (45.7%), although more instances of idiopathic isolated unilateral HNP did not complain of symptoms or had no symptoms documented (50%) than whole HNP cases (22.7 percent).[23] While prodromal symptoms were related with the main illness in whole HNP patients, idiopathic unilateral HNP tended to emerge abruptly without prodromal symptoms. Dysarthria (58.3%), dysarthria with dysphagia (20.8%), tongue weakness and leathery sensation (8.3%), and trouble chewing food(8.3 percent) were the most prevalent early symptoms. 70.8 percent exhibited tongue deviation, 50 percent had fasciculation, 45.8% had tongue atrophy, and 29.2% had tongue weakness on physical examination. Unilateral paresis generally produces weakening and wasting of the ipsilateral tongue, with minor dysarthria and dysphagia due to effective compensation by the contralateral muscles. There were no early clinical signs of a favourable prognosis (e.g., absence of fasciculation or lack or rapid resolution of pain).[20] There were no prodromal signs in our patients, and none of them complained of severe dysarthria or dysphagia. Hematological and biochemical blood tests revealed that all of the patients were in good health. In 62.5 percent of the patients, negative viral serology supported these findings. It has previously been explored the link between isolated HNP and viral infection, such as infectious mononucleosis, the common cold, and influenza vaccine. However, isolated HNP in combination with viral infection has been described in just a few investigations over the last decade. The cause of solitary HNP instances involving viral infection is unknown.[25] A total of 20 individuals had CT scans, and all of them were found to be normal. Twenty-two patients had further imaging (MRI of the craniofacial region with or without contrast/MRI of the neck) to evaluate the hypoglossal canal and nerve, but no abnormalities, tumour, or lesion was seen. Tongue atrophy, or fatty replacement of the tongue muscle, accounted for 16 percent of abnormalities on MRI. In instances of HNP, thorough neuroimaging examinations are required to rule out tumoral, spontaneous, traumatic, or vascular causes. To rule out sarcoidosis, a chest radiograph should be taken. A space-occupying lesion along the hypoglossal nerve's route may be discovered on CT scans of the head and neck. MRI is very good for obtaining soft tissue features and may reveal hypoglossal nerve demyelination. Furthermore, these imaging techniques will enable for the detection of vascular abnormalities, which may be useful for determining the need for particular or early anticoagulant medication.[3] There was no particular therapy for idiopathic isolated unilateral HNP in the majority of instances (16 patients; 66.7 percent). Four patients were given oral steroids or multivitamins, whereas five patients had no therapy. In terms of prognosis, 15 patients (62.5 percent) demonstrated progressive improvement and full resolution after a year, whereas nine patients (37.5 percent) did not. Almost all of the patients made a full recovery without any therapy, and some researchers feel this is a clinical entity comparable to Bell's palsy.[6] Our patients were treated with steroids in accordance with the Bell's palsy treatment protocol.[26] Therapy, on the other hand, is debatable, and there was no link between treatment and prognosis. Surprisingly, individuals with an illness duration less than the median (7.5 days) had a favourable outcome. Patients with an illness duration of

up to 7.5 days (12 patients) recovered completely, whereas those with a disease duration of more than 7.5 days (3 patients) recovered completely or did not recover at all (9 patients). These findings suggest that idiopathic isolated unilateral HNP with a short duration has excellent outcomes. This might be due to the fact that the symptoms and functionality of idiopathic HNP alter with time.[25]Furthermore, all patients with a dismal prognosis had tongue atrophy, with adipose tissue replacing tongue muscle. As a result, we propose MRI confirmation of the tongue status for prognosis calculation. To our knowledge, this is the first comprehensive assessment of the literature for idiopathic isolated unilateral HNP. Although this illness is extremely rare, it should be treated with caution. While the majority of instances, such as Bell's palsy, demonstrate satisfactory healing without any special therapy, it should be noted that over 50% of cases have permanent effects. Finally, because HNP is such a complicated illness, an in-depth and step-by-step approach to etiological diagnosis is required.

CONCLUSION

In these cases, the significance of a thorough and rigorous work-up cannot be overstated in order to rule out the more prevalent curable and reversible illnesses. In chronic instances, it is suggested that long-term follow-up be started. In the absence of any additional neurological complaints, Ho et al urge a complete neurological evaluation and a repeat MRI scan every 3–5 years.[7]

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Conflict of interest

There are no conflict of interest.

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