Idiopathic Vocal Cord Palsies and Associated Neurological Conditions

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**Objective:** To retrospectively review the clinical case records of patients with idiopathic vocal cord palsies (VCPs) for the presence of preexisting or subsequent development of neurological disease, including multiple sclerosis, motor neuron disease, myasthenia gravis, cerebrovascular disease, and Guillain-Barré syndrome.

**Design:** Retrospective case review of all patients with VCP presenting sequentially within a 45-month time span.

**Setting:** Tertiary referral center.

**Patients:** One hundred ninety-three patients with VCP.

**Results:** Thirty-five cases of VCP (18.1%) were idiopathic. Eight (22.8%) resolved after a mean time of 5 months. A preexisting central nervous system condition was noted in 9 (25.7%) of 35 patients with idiopathic VCP. A subsequent central nervous system condition developed in 7 patients (20.0%). These included 2 cases of cerebrovascular accidents, 1 case of postpolio syndrome with respiratory failure, and 1 case of polyneuropathy secondary to paraneoplastic syndrome.

**Conclusions:** A high frequency of neurological conditions was observed in adult patients initially presenting with idiopathic VCP. Patients with VCP but without overt neurological disease may also subsequently develop a serious neurological condition. Careful neurological evaluation of all patients with idiopathic VCP is recommended.

racic/cardiac surgical procedures were nearly equal causes of VCP (n=33). Forty-six cases (23.8%) were due to tumors, 19 (9.8%) to trauma, 17 (8.8%) to cerebrovascular disease, and 7 (3.6%) to congenital and inflammatory conditions. Thirty-five cases (18.1%) were idiopathic (Table 1).

There were nearly equal numbers of men (n=18) and women (n=17) with idiopathic VCP. Mean age was 63.5 years (age range, 22-87 years). The mean follow-up period was 23¼ months (range, 0-116 months). All cases were unilateral. More than twice as many cases of idiopathic VCP were found on the left side as on the right side (24 vs 11). A preexisting central nervous system condition was noted in 9 (25.7%) of 35 patients. A subsequent central nervous system condition developed in 7 patients (20.0%) with a mean time to diagnosis of 17 months (range, 2 weeks to 4 years). Preexisting and subsequently developed central nervous system conditions identified are listed in Table 2. Eight cases (22.8%) of idiopathic VCP resolved in a mean time of 5 months (range, 23 days to 26 months).

**COMMENT**

In this series of adult patients, nearly half of the patients with idiopathic unilateral VCP presented with, or were subsequently diagnosed as having, neurological conditions. Our study confirms and extends evidence for such an association (Table 3).

Several retrospective reviews have suggested an association in children between idiopathic VCP and underlying neurological conditions. In a consecutive sample of 102 cases of pediatric VCP, 35% were classified as idiopathic and 16% as having underlying neurologic conditions. Seven patients had Arnold Chiari malformation, 2 exhibited peripheral neurological disease, 1 had hereditary distal spinal muscular degeneration, and 1 had Horner syndrome ipsilateral to the VCP. Of 113 children diagnosed with congenital VCP, excluding postsurgical cases, 37% had idiopathic VCP, and 25% associated neonatal neurological diseases.

In adults, the rate of idiopathic VCP is similarly high and is sometimes associated with a neurological condition (Table 3). In a series of 61 adult patients with bilateral...
eral VCP, 39% of cases were idiopathic. However, in that series, despite detailed ear, nose, and throat, neurological, and radiological examinations, no underlying neurological disorders were disclosed. Havas et al classified etiologies of unilateral VCP in 108 patients. Forty-five cases were iatrogenic, and 36 cases were idiopathic. Of the remaining 27 patients, 6 had a central nervous system disorder or systemic neurological disorder, 3 had vagus neuroma or neurofibroma, and 2 had postpolio syndrome. Ramadan et al evaluated 98 patients with unilateral VCP. The cause was found to be neoplastic disease in 32% of the cases; idiopathic in 16%; or caused by surgery (30%), trauma (11%), central nervous system disorder (8%); all 8 cases were the result of stroke), or infection (3%). Associated cranial nerve injuries were found in 15 patients. Five patients had injuries to cranial nerve 11, and 4 patients, to cranial nerve 12. Srisrompotong et al evaluated 90 patients with unilateral VCP, finding that 29% of cases were due to neoplasm; 21%, inflammation; 8%, trauma from endotracheal intubation and external laryngeal trauma; and 5%, central nervous system disease. Twenty-four percent were iatrogenic, and 13% were idiopathic.

PATHOPHYSIOLOGIC CONSIDERATIONS

In our series of patients with unilateral VCP, it is difficult to make a pathophysiological connection between idiopathic VCP and some diagnoses (eg, dementia or presyncope) (Table 2). How and why should a patient with multiple cranial neuropathies or postpolio syndrome be prone to develop VCP? There are isolated forms of idiopathic Guillain-Barre-type illnesses, inflammatory or infiltrative neoplastic skull base diseases, and a variety of other similar disorders that could logically affect both cranial nerves and their various branches. The neurological conditions that preceded VCP (in 5 of 9 cases) seem to be associated with either the brainstem outflow end of the neuraxis or the segmental laryngeal or esophageal anatomy (the neuromuscular disorders, multiple cranial neuropathies, esophageal dysmotility, and laryngeal spasms).

Similarly, it is unclear whether VCP potentially heralds any subsequent neurological diagnosis. The subsequent neurological conditions included hearing loss or vertigo and postpolio neuropathies. These conditions seem to suggest an association of idiopathic VCP with neuromuscular disorders because 9 (26%) of 35 patients have either a neuromuscular or closely aligned otolaryngologic diagnosis before or after VCP is diagnosed. The left vocal cord is more vulnerable to injury than the right, as noted herein. The left recurrent laryngeal nerve is longer. Typically, there is a 28% difference in length; it can vary from 5 to 15 cm. In addition, there is pronounced variation in the way in which the 2 recurrent laryngeal nerves meet the larynx. In a now-discontinued procedure for the treatment of spasmodic dysphonia, the left recurrent laryngeal nerve was chosen for sectioning more often than the right because the right has a lower risk of subsequent disease. Whether such anatomical differences and susceptibility to disease are evidenced by the 2-fold increased incidence of left-sided VCP in our study remains to be proved.

CENTRAL NERVOUS SYSTEM DISORDERS VS PERIPHERAL NEUROLOGICAL ETIOLOGIES

The neurological disorders reported to be associated with VCP seem to be divisible into central nervous system disorders (cerebrovascular disease, multiple sclerosis, multiple system atrophy, Parkinson disease, progressive supranuclear palsy, and Arnold-Chiari malformation) and peripheral nervous system disorders (eg, Wallenberg syndrome, myasthenia gravis, Guillain-Barré syndrome, postpolio syndrome, congenital hypomyelination neuropathy, Charcot-Marie-Tooth disease, and chronic motor axonal neuropathy). Although cases of idiopathic VCP can occasionally be bilateral, they are usually unilateral. Our series of patients were all adults with unilateral VCP, and nearly half (46%) either had preexisting or subsequently developed comorbid neurological conditions.

Our study validates findings that cerebrovascular disease is a common cause of VCP. Seventeen (9%) of the 69 cases reviewed were caused by stroke. Two patients previously diagnosed as having idiopathic VCP, 1 of whom had a history of transient ischemic attacks, subsequently experienced cerebrovascular accidents.

Two of our 35 patients with idiopathic VCP, 1 of whom subsequently developed respiratory failure, were diagnosed as having postpolio syndrome. Common signs and symptoms include breathing or swallowing problems and sleep-related breathing disorders. Postpolio laryngeal muscle weakness requiring surgical intervention is reported.

LIMITATIONS

In our case series, we were unable to perform extensive medical chart abstractions on an age-matched control group that would enable us to determine rates of the identified neurological conditions in our general population for comparison with the rates seen among our patients with VCP. In addition, the mean time elapsed from diagnosis of VCP to a subsequent neurologic condition in our patients was 17 months (range, 2 weeks to 4 years). Likewise, the time between the diagnosis of a preexisting neurological condition and the subsequent evolution of VCP was not determined. Therefore, any cause-and-effect relationships between neurological conditions and idiopathic VCP cannot be established. Rather, our findings suggest a need for a further prospective cohort study of patients with idiopathic VCP with comparison to an age-matched, population-based control group to ascertain whether a valid epidemiological and temporal association exists between idiopathic VCP and neurological disorders.

In conclusion, the number of patients diagnosed with VCP is rising as physicians become more aware of its prevalence. Given the frequency of associated neurological conditions in adult patients with idiopathic VCP, a careful neurological examination should be considered for all patients. A prospective study focusing on a serial neurological evaluation in patients with idiopathic VCP should be mounted.
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References:


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