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Collet–Sicard syndrome caused by a paraganglioma in the region of the jugular foramen on one side: A case report and review

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Abstract

Collet–Sicard syndrome is a rare neurological disorder caused by injury to the cranial nerve pairs IX, X, X, and XII. The author reports on a previously fit 27-year-old man who presented with dysphagia, choking on drinking water, hoarseness, weakness when turning the neck and shrugging the shoulders, and unexplained weight loss. Enhanced magnetic resonance imaging indicated a space-occupying lesion at the right jugular foramen. After surgical resection, the pathologic findings suggested a paraganglioma of the right jugular foramen and confirmed the diagnosis of Collet–Sicard syndrome. After postoperative treatment with a combination of acupuncture and modern medicine, the patient's symptoms significantly improved. This article analyzes previous literature regarding Collet–Sicard syndrome etiology and reports the case of a patient with a rare etiology, whose prognosis improved significantly after treatment with a combination of acupuncture and modern medicine.

Keywords

Collet–Sicard syndrome, cranial nerve palsy, jugular foramen, hypoglossal canal, paraganglioma, case report

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Introduction

Collet–Sicard syndrome (CSS), also known as occipital condyle-jugular foramen junction syndrome and hemilaryngeal–pharyngeal–shoulder–pharyngeal paralysis syndrome, is a very rare disorder caused by a skull base lesion affecting the jugular foramen and hypoglossal canal or a lesion extending from the jugular foramen to the canal of the anterior occipital condyle, resulting in the peripheral paralysis of four pairs of lower cranial nerves (the glossopharyngeal, vagus, accessory, and hypoglossal nerves). This paralysis manifests as hoarseness of the voice, difficulty swallowing, impairment of taste in the posterior third of the tongue or paralysis of the soft palate of the ipsilateral side, and paresthesia and atrophy of the lingual, sternocleidomastoid, and trapezius muscles. The causes of CSS include tumors (primary or metastatic), trauma, vascular lesions, inflammation, and medical complications. According to previous studies,[1] invasive metastatic processes, skull base fractures, and carotid artery dissection are the most common etiologies. Primary intracranial tumors are an extremely rare cause of CSS. The diagnostic aspects of all reported cases of primary intracranial tumors causing CSS include hoarseness, dysphagia, paralysis of the soft palate or uvula, and paralysis and atrophy of the lingual, sternocleidomastoid, and trapezius muscles. Depending on the location of the tumor or the extent of the lesion, headache,[2,3] vomiting,[2,3] vertigo,[4,5] tinnitus or impaired hearing,[4,6] facial paralysis or decreased facial sensation,[4] and unsteady gait[4] may also be observed. An occupancy centered in the jugular foramen area and extending to the periphery is observed on

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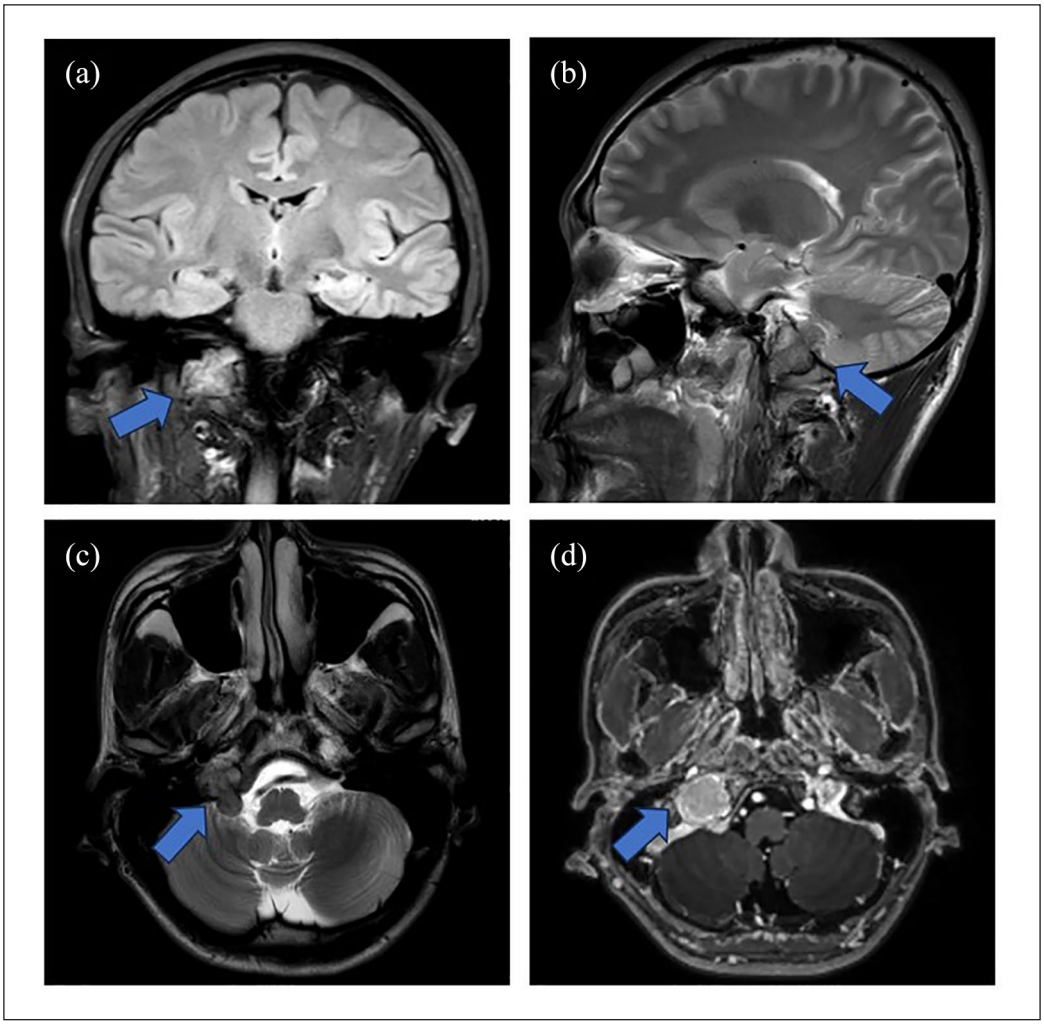

Figure 1. FLAIR (fluid attenuated inversion recovery) images (a) demonstrated a high signal in the right jugular foramen area (blue arrow); T2-weighted images (b) and (c) demonstrated moderate signal intensity in the right jugular foramen area (blue arrows); coronal 26 mm × 22 mm (a), sagittal 20 mm × 26 mm (b), and horizontal 26 mm × 22 mm (c). Enhanced magnetic resonance imaging revealed a slightly longer T2 signal mass in the right jugular foramen area (d).

imaging, with a low signal on T1-weighted images and a moderate or high signal on T2-weighted images. Clinical diagnostic tests, such as a video fluoroscopic swallowing study (VFSS) to assess dysphagia, laryngoscopy for vocal cord paralysis, staining for pyriform fossa remnants, and trapezius and sternocleidomastoid electromyography can be used as auxiliary diagnostic tools; however, the gold standard for clinical confirmation is pathologic tissue biopsy.

Defining the cause of CSS is important for its diagnosis and treatment. The author's literature search revealed that most case reports do not summarize the disease etiology, and no effective treatment plan has been proposed to improve CSS patient prognosis. This article summarizes CSS etiology and, more importantly, reports the case of a CSS patient with a right jugular foramen paraganglioma whose symptoms improved significantly after treatment with acupuncture combined with modern medicine.

Case description

A 27-year-old man was admitted to the hospital on July 11, 2023, with the complaint of "dysphagia and choking on water for more than 1 year." The patient presented with dysphagia and choking on drinking water in 2022, and the symptoms worsened progressively over 1 year. On May 18, 2023, during a physical examination for worsening symptoms, such as difficulty eating and drinking and progressive weight loss, cranial and enhanced magnetic resonance imaging (MRI) analyses were performed, indicating an occupation in the right jugular foramen region with a high possibility of jugular vein bulboma (Figure 1(a)–(d)). In June 2023, the patient presented with hoarseness, difficulty swallowing, choking on water, coughing up sputum, and weight loss. There was no history of hypertension, diabetes mellitus, or coronary heart disease. Due to the lack of relevant treatment in the past, to address the intracranial occupying etiology, the

patient underwent a right posterior curved far lateral craniotomy of the right ear under general anesthesia with reconstruction of the skull base and removal of the adipose fascia on June 27, 2023, and the operation went smoothly. The postoperative pathological indications combined with morphological and immunohistochemical findings were consistent with paraganglioma with severe partial extrusion and deformation, CNS WHO grade 1, and focal infiltration of bone tissue. The immunohistochemistry results were as follows: CK (−), CgA (+), Syn (+), S100 (scattered), Ki-67 (+, 2–5%), SSTR-2 (+), and PR (scattered +). Dysphagia, hoarseness, choking on drinking water, coughing and phlegm symptoms, and a nasal diet persisted after the surgery.

On examination, an arc-shaped surgical scar approximately 12 cm in length was observed behind the right ear. The patient's tongue extension was slight to the right, with atrophy of the right lingual muscle and an uneven tongue surface. His hearing was normal. The uvula was deviated to the left, and the elevation of the soft palate on the right side was limited. The pharyngeal reflex was weakened, and the right side of the neck was slightly weaker. The right sternocleidomastoid muscle was atrophied and weak, and the right side of the shrug was weaker than the left side (Supplemental material Figure S1(a)–(d)). The VFSS results revealed dysphagia (pharyngeal and esophageal phases), overt aspiration, and loss retardation of the cricopharyngeal muscle (Supplemental material Figure S2(a) and (b)). Electron laryngoscopy revealed a fixed paracentral position of the right vocal folds with limited movement, limited left vocal fold movement, poor bilateral vocal fold closure, and a large volume of foamy secretion trapped in the hypopharynx, predominantly in the right pyriform fossa; swallowing of the stain showed residue in the right pyriform fossa (Supplemental material Figure S3(a) and (b)). The current diagnosis is CSS.

The author treated the patient with acupuncture combined with modern medicine. Lianquan (CV23), Jialianquan, and the suprahyoid muscle group were selected for acupuncture, and electroacupuncture (EA) was applied at Jialianquan with a discontinuous wave, a frequency of 5 Hz, and a current intensity of 1 mA. EA was applied once daily for 30 min (Supplemental material Figure S4(a)). Regarding rehabilitation therapy, low-frequency pulse electrical stimulation was applied to the hyoid muscle group once daily for 20 min at a stimulation intensity tolerable for the patient (Supplemental material Figure S4(b)). Moreover, daily rehabilitation training, including swallowing function training, vocal training, and compression and stretching of the sternocleidomastoid muscle, was performed. Regarding medication, an ultrasound-guided botulinum toxin A injection was given to improve spasm in the cricopharyngeal muscle (Supplemental material Figure S4(c)), together with a methylcobalamin intramuscular injection and oral vitamin B1 tablets to nourish the nerves.

After 6 months of treatment, a physical examination revealed that the deviation of the tongue extension and atrophy of the lingual muscle had improved. The patient's atrophy of the right sternocleidomastoid muscle also improved, and his neck turning and shoulder shrugging strength increased (Supplemental material Figure S5(a)–(d)). The VFSS assessment showed no aspiration, the cricopharyngeal muscles were mostly open, and there was a slight residue in the right pyriform fossa (Supplemental material Figure S6(a) and (b)). The patient no longer requires a nasogastric tube, and his weight has significantly increased.

Discussion

CSS is a very rare condition caused by a lesion at the skull base affecting the jugular foramen and hypoglossal canal or a lesion extending from the jugular foramen to the canal of the anterior occipital condyle, resulting in peripheral paralysis of four pairs of lower cranial nerves (the glossopharyngeal, vagus, accessory, and hypoglossal nerves). CSS paralysis manifests as hoarseness of the voice, dysphagia, impairment of taste in the posterior third of the tongue or paralysis of the ipsilateral soft palate, and palsy and atrophy of the lingual, sternocleidomastoid, and trapezius muscles. Causes of CSS include tumors (primary or metastatic), trauma, vascular lesions, inflammatory processes, and medical complications. The hypoglossal canal is a bony foramen located at the anterolateral margin of the foramen magnum of the occipital bone (in the middle and posterior thirds of the occipital condyle); the hypoglossal nerve passes through this foramen, which is closely adjacent to the posterior medial aspect of the jugular foramen. The hypoglossal nerve is closely associated with the three nerves of the jugular foramen; thus, the hypoglossal nerve is susceptible to combined damage with these three nerves.

We searched and screened the literature in PubMed related to the onset of CSS between 1961 and 2022. The search terms "Collet–Sicard syndrome" and "Collet Sicard" were used, and 100 reports were screened; etiologies were counted independently in cases of CSS with multiple etiologies. The data are detailed in Supplemental material Table S1.

Among the cases reported in the 100 publications, 19 patients had CSS due to traumatic factors, accounting for 19% of the total, with head trauma and Jefferson fracture predominating. CSS due to vascular factors accounted for 22 (22%) patients (22% overall), with internal carotid artery entrapment being the most common. A total of 43 cases of CSS due to tumor factors accounted for 43% of the total number of cases, 20 of which were due to malignant tumor metastases, accounting for 46.5% of CSS caused by tumors. CSS due to prostate cancer had the highest reporting frequency. Laigle-Donadey et al.[7] suggested that the most common skull base metastases are prostate and breast cancers; lung cancer is the fourth most common type of cancer, accounting for approximately 6% of all skull base metastasis cases. Rhiannon et al.[8] performed a total of 5,644 imaging examinations, including of the brain, on 4,341 Australians with prostate cancer. Eight of these patients were diagnosed with concurrent intracranial metastases, demonstrating the

low incidence of intracranial metastases in prostate cancer patients (0.18%). In recent years, studies on the incidence of intracranial metastases from prostate cancer have been rare, and there is little evidence to suggest that prostate cancer is associated with an increased risk of complicating intracranial metastases. A total of 16 cases of CSS were caused by primary benign intracranial tumors, accounting for 37.2% cases caused by tumors; 2 of these cases were caused by paragangliomas,[9,10] accounting for 12.5% of cases arising from benign tumors. The case reported in this article is in the benign tumor etiology category.

Invasive metastasis of tumors, vascular lesions such as internal carotid artery entrapment, and head trauma are common causes of CSS; the number of cases arising from primary intracranial tumors is relatively small. Of the 51 cases reports on CSS etiology from 1915 to 2012, only two were secondary to jugular vein bulbomas, and one was secondary to a nerve sheath tumor of the hypoglossal nerve.[1] Reports on primary intracranial tumors leading to CSS have steadily increased, but this causative factor remains a relatively rare CSS etiology.

Notably, the syndromes resulting from multiple cranial nerve injuries in tumor category are determined by the number of cranial nerves involved and not by the duration of the disease, that is, by space rather than time.

In our present case, CSS was clearly defined as a lesion caused by a paraganglioma, a benign tumor in the jugular foramen region. This rare etiology was determined according to the patient's preoperative cranial MRI, clinical presentation, and postoperative immunohistochemical biopsy. At the onset of the disease, the tumor in the jugular foramen area compressed cranial nerves IX, X, and XI, which run through the jugular venous foramen, resulting in choking on drinking water, difficulty swallowing, dysarthria, weakening of the pharyngeal reflexes, weak turning of the neck, atrophy and weakness of the sternocleidomastoid muscles, and weakness when shrugging the shoulders. The patient's disease started in the area of the jugular foramen, and the illness duration was long, with progressive weight loss; therefore, the disease may have progressed between the patient's first presentation and diagnosis. The paraganglioma was considered to have progressed in size, and the lesion compressed the XII cranial nerve in the hypoglossal canal below it, resulting in right deviation of the tongue extension and atrophy of the right lingual muscle.

CSS is primarily addressed by treating the cause of the symptoms, and rehabilitation training should be conducted as soon as possible after the cause of the disease is removed. Busto-Crespo et al.[11] found that rehabilitation started within 1 year of the onset of vocal cord paralysis was better than rehabilitation later than 1 year after onset; however, treatment later than 1 year after onset can still partially improve the patients' hoarseness symptoms and quality of life. A clinical study by Zhang Ran et al.[12] noted that early, comprehensive rehabilitation in patients with vocal cord paralysis combined with parasympathetic nerve injury could improve rehabilitation progress. Patients with CSS have more complications from primary lesions combined with posterior cranial nerve injuries, and early and comprehensive rehabilitation training is needed to address their dysfunction.

In recent years, several meta-analyses[13–15] have revealed that acupuncture combined with rehabilitation training, electrical stimulation, and other modalities for patients with dysphagia can achieve significant therapeutic efficacy, better than that of rehabilitation training, physical, or chemical intervention alone. Relevant dysphagia rehabilitation guidelines[16,17] strongly recommend acupuncture, surface neuromuscular electrical stimulation combined with conventional swallowing training, motor behavior therapy, and botulinum toxin injections for improving cricopharyngeal muscle dysfunction, supporting the choice of this treatment modality. The above literature provides the basis for our choice of treatment modality.

In the present case, rehabilitation exercises, such as swallowing function training, vocal training, and compression and stretching of the sternocleidomastoid muscle, were combined with acupuncture and low-frequency pulse electrical stimulation. Lianquan (CV23), Jialianquan, and the suprahyoid muscle group were selected for acupuncture, and Jialianquan was treated with EA. Da et al.[18] revealed the basis of using the neural circuits of acupuncture Lianquan (CV23) and mylohyoid with EA stimulation to improve dysphagia through animal experiments, indirectly confirming the effectiveness of stimulating this site. In a randomized controlled clinical trial, Jin et al.[19] demonstrated that EA at the suprahyoid muscle group could improve dysphagia in patients with pharyngeal phase swallowing disorders more effectively than conventional acupuncture. The main mechanism of EA at this site is to increase the mobility of the hyoid laryngeal complex to improve swallowing disorders.

In this case, the treatment of EA combined with modern medicine brought significant curative effect to the patient. Since CSS is rare, with various causes and multiple symptoms, a single case report does not show that any treatment, such as EA, low-frequency pulse electrical stimulation, rehabilitation training or botulinum toxin injection, can produce significant effects on the disease alone. We believe that the patient benefited from the treatment reported in this case, which may be due to the combined effect of acupuncture and modern medical treatment. This provides a new idea for clinicians to make treatment decisions in the future, and whether any treatment method can produce curative effect still needs further clinical research.

Conclusion

CSS is a rare neurological disorder, and this article reports the case of a patient with CSS caused by a paraganglioma in the jugular foramen region, which is a relatively rare etiology. The etiology of CSS was classified and summarized by compiling the previous literature to enable more clinicians to better recognize this rare disease and develop more ideas for clinical diagnosis. More importantly, this patient achieved a

better prognosis through the combination of acupuncture and modern medicine, providing a unique approach to the clinical treatment of CSS.

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Author contributions

Y.H. Conceptualization; writing—original draft; writing—review and editing; Y.H. Investigation; writing—review & editing; Y.M. Investigation; writing—review & editing; H.J. Conceptualization; visualization; funding acquisition.

Availability of data and materials

The authors have full access to the patient data.

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Ethics approval

Ethics committee approval was not required because the article is a retrospective description of a single patient.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

Consent for publication

Written informed consent documentation was obtained from the patient, who voluntarily consented to the use of his case and video recording of his abnormalities for education purposes, including publication in a journal, without compensation.

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Supplemental material

Supplemental material for this article is available online.

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