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The Spine Journal ■■ (2016) ■■–■■

THE SPINE JOURNAL

Case Report

Developmental abnormalities of the craniocervical junction resulting in Collet-Sicard syndrome

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Received 23 July 2015; revised 20 April 2016; accepted 28 April 2016

Abstract

BACKGROUND CONTEXT: Collet-Sicard syndrome describes the paralysis of cranial nerves IX–XII and is the most frequently reported neurologic complication associated with Jefferson fractures. As the lateral mass of the atlas is displaced laterally toward the styloid process and the stylohyoid ligament, the lateral mass impinges on cranial nerves IX–XII. However, Collet-Sicard syndrome in association with other anomalies of the atlas has rarely been reported.
PURPOSE: The aim of this study was to report an unusual case of Collet-Sicard syndrome as a result of developmental abnormalities of the craniocervical junction.
STUDY DESIGN/SETTING: This is a case report of a single patient.
METHODS: Chart and radiographic data were reviewed and reported.
RESULTS: We report a 70-year-old man who developed hoarseness, dysarthria, and dysphagia from developmental abnormalities of the craniocervical junction including a congenital occiput–C1–C3 fusion and hypoplastic dens. On computed tomography, the distance between the left transverse process of the atlas and the left styloid process of the skull was 3 mm.
CONCLUSION: In suspected Collet-Sicard syndrome, developmental abnormalities of the craniocervical junction should be considered in the differential diagnosis.

Keywords: Assimilation of the atlas; Collet-Sicard syndrome; Cranial neuropathy; Craniocervical junction; Developmental disorders; Hypoplastic dens

Introduction

Collet-Sicard syndrome describes the paralysis of cranial nerves IX–XII [1]. It is usually caused by a posterior lacerocondylar space lesion, where these nerves are closely related [2]. Earlier reports in the literature describe Collet-Sicard syndrome as being caused by skull base tumors of primary and metastatic origin (breast, prostate, lung, renal cell, and cervix) [3–6], multiple myeloma [7], dissection and coiling of the internal carotid artery [8,9], vasculitic conditions such as polyarteritis nodosa [10], jugular veins thrombosis [2], closed head injury [11], skull base fractures [12], and penetrating trauma [13]. In addition, Collet-Sicard syndrome is the most frequently reported neurologic complication associated with Jefferson (C1) fractures [1,14,15]. As the lateral mass of the atlas is displaced laterally away from the spinal cord toward the styloid process and the stylohyoid ligament, the lateral mass impinges on cranial nerves IX–XII [1,14,15]. Damage to the cranial nerve IX (glossopharyngeal nerve) causes loss of taste in the posterior tongue and the ipsilateral gag reflex and poor pharyngeal elevation with swallowing and speaking, resulting in dysphagia and dysarthria [1,16]. Impairment of the cranial nerve X (vagus nerve) causes dysphagia, hoarse speech, and ipsilateral paralysis of the soft palate, and loss of laryngeal and pharyngeal sensation [1,16]. The soft palate droops on the ipsilateral side and deviates to the contralateral side during phonation [17]. Direct laryngoscopy can show ipsilateral paralysis of the vocal cords [16]. Damage to cranial nerve XI (accessory nerve) causes paralysis of the sternomastoid and the upper fibers of the trapezius [1,17]. Impairment of the cranial nerve XII (the hypoglossal nerve) results in paresis and atrophy of half of

FDA device/drug status: Not applicable.

Author disclosures: ***KK:*** Nothing to disclose. ***BGM:*** Nothing to disclose.

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http://dx.doi.org/10.1016/j.spinee.2016.04.022
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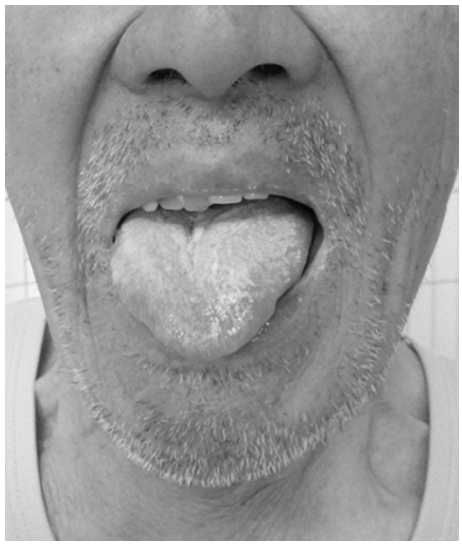

Fig. 1. Photograph of the patient. His tongue deviates to the left.

the tongue [16,17]. On protrusion, the tongue deviates to the affected side [16,17].

However, Collet-Sicard syndrome in association with other anomalies of the atlas has rarely been reported. In this report, we discuss a patient whose only initial presentation of developmental abnormalities of the craniocervical junction was most akin to a Collet-Sicard syndrome.

Case report

A 70-year-old man with a history of limited neck motion, hypertension, and left facial palsy presented to our hospital with 2 weeks of difficulty articulating words and 10 days of dysphagia to solids. He had limitation of rotary motion of his neck after trauma that happened several decades before. There was no history of recent trauma. He had a left lower motor facial nerve palsy, a decreased gag reflex on the left, the soft palate pulling to the right, and weakness of left trapezius and sternocleidomastoid. His tongue deviated to the left (Fig. 1). He had no Horner's syndrome. Cervical spine range of motion was restricted in rotation bilaterally, but he did not have neck pain. Direct laryngoscopy revealed left vocal cord paralysis. The overall clinical picture was consistent with dysfunction of the cranial nerves VII (a previous Bell's palsy), and IX–XII (a Collet-Sicard syndrome). X-ray (Fig. 2), computed tomography (CT) (Fig. 3) and magnetic resonance imaging (Fig. 4) of cervical spine showed a congenital occiput–C1–C3 fusion, hypoplastic dens, and scoliosis. The left transverse processes of the atlas and axis were hypertrophic (Figs. 2, Left, and 3, Left). On CT, the distance between the styloid process of the skull and atlas transverse process was 3 mm left and 13 mm right (Fig. 3, Right). Brain diffusion-weighted magnetic resonance imaging was normal, and brain and neck magnetic resonance angiography (Fig. 5) showed absent right vertebral artery flow and the elongated and stenotic basilar artery. Rheumatoid factor assay was 10.7 IU/mL (normal range, 0–18 IU/mL). He had swallowing abnormalities shown by videofluoroscopic modified barium swallow study. Delayed swallow, residual barium in the vallecula and piriform sinuses, and aspiration were noted during the videofluoroscopic modified barium swallow study. He was prescribed a dysphagia diet and received swallowing therapy such as oral exercises, methods of postural facilitation, and transcutaneous neuromuscular electrical stimulation. He was discharged 8 days after admission and refused recommended follow-up, seeking a second opinion from another physician.

Discussion

Occipitalization or assimilation of the atlas refers to congenital partial or complete fusion of the atlas to the occiput [18,19]. Unilateral occipitalization of the atlas can cause torticollis in young children [19,20]. There can be other anomalies, such as congenital fusion of the second and third cervical vertebrae [18–20]. Atlantoaxial instability may result from aplasia or hypoplasia of the odontoid process or with

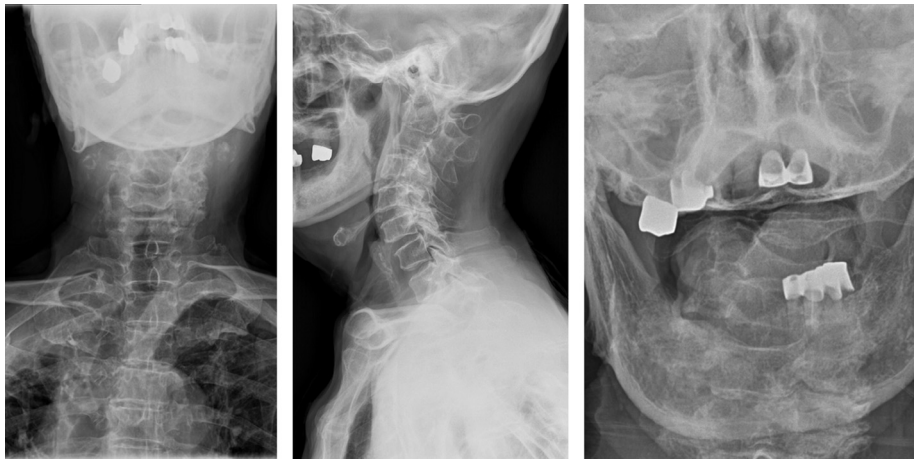

Fig. 2. Cervical spine radiographs. Anteroposterior (Left), lateral (Middle), and open-mouth odontoid (Right) radiographs show an occiput–C1–C3 fusion, hypoplastic dens, and scoliosis.

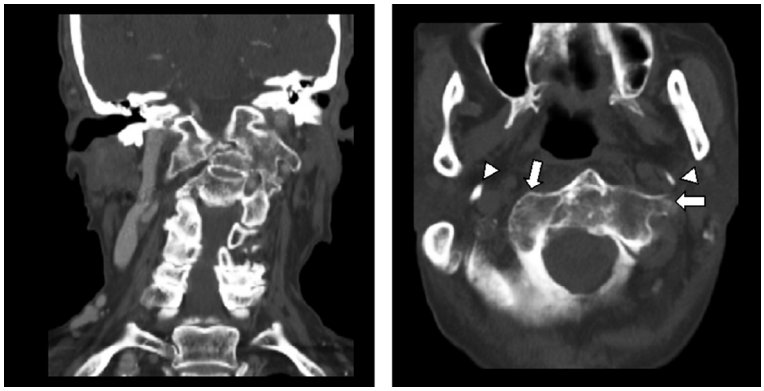

Fig. 3. Computed tomography of the neck. (Left) Computed tomography scan coronal view, bone window, shows an occiput–C1–C3 fusion, hypoplastic dens, and scoliosis. (Right) Computed tomography scan axial view, bone window, demonstrates that the distance from the styloid process (arrowhead) to the atlas transverse process (arrow) was 3 mm left and 13 mm right.

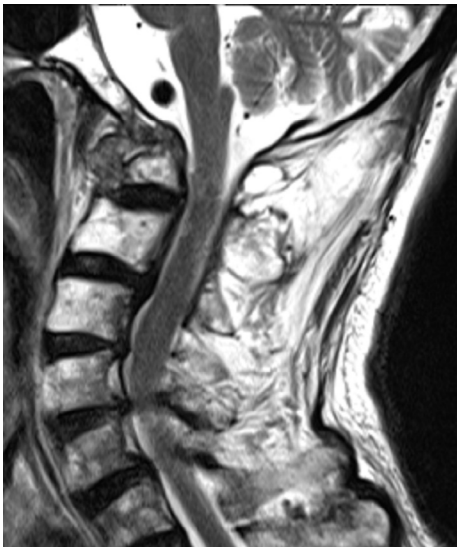

Fig. 4. Magnetic resonance imaging of the neck. A sagittal T2-weighted magnetic resonance image shows a hypoplastic odontoid process.

assimilation of the atlas [18–20]. The articulation between the first and second cervical vertebrae is the most mobile part in the vertebral column and is the most inherently unstable [19]. The odontoid process acts as a bony buttress that prevents hyperextension [19]. However, the remainder of the normal range of motion is maintained and depends on the integrity of the capsular and ligamentous structures [19]. When transverse atlantal ligament is ruptured, a relative ventral shift of the atlas over the axis can result in brainstem injury by impingement against the intact odontoid process [19]. If the odontoid process is absent, the risk of brainstem injury is low [19]. In addition, chronic atlantoaxial dislocation may provoke the formation of granulation tissue that can constrict the cervicomedullary junction, resulting in neurologic deficit [19,20]. Symptoms associated with vertebral artery compression may occur [19,20]. Management of assimilation of the atlas is complex and needs to be individualized [21]. Nonoperative methods such as immobilization in cervical orthosis should be attempted at first in patients who have minor

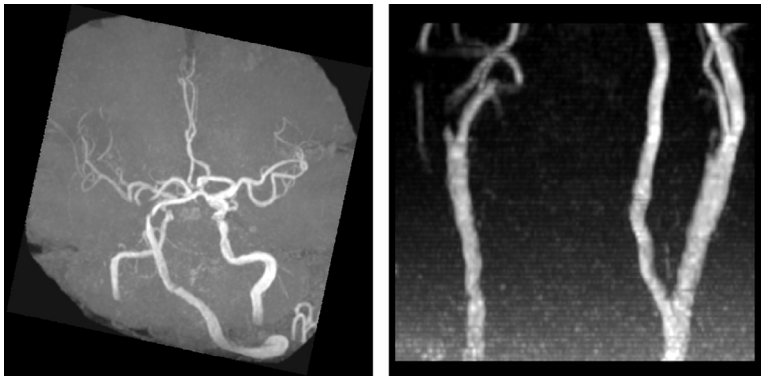

Fig. 5. Brain and neck magnetic resonance angiography shows absent right vertebral artery flow and the elongated and stenotic basilar artery.

symptoms resulting from assimilation of the atlas [21]. When signs and symptoms are caused by anterior impingement, occipital cervical fusion is indicated [21]. When signs and symptoms are caused by posterior impingement, posterior decompression and occipital cervical stabilization are indicated [21]. However, cranial neuropathy in our patient was not due to vascular compromise of the vertebral artery nor to compression of the brainstem from the backward-projecting odontoid or retro-odontoid pannus.

Collet-Sicard syndrome refers to the rare finding of unilateral lesions of cranial nerves IX–XII [1,14]. Cranial nerves IX–XI exit the skull through the jugular foramen; cranial nerve XII exits through the hypoglossal foramen. After exiting the skull, the cranial nerves pass through the space between the styloid process and the transverse process of the atlas [1,14]. There is normally 8–10 mm of space between them [15]. In our patient, the distance from the styloid process to the transverse process of the atlas was only 3 mm on the left (Fig. 3, Right). We propose that as the left transverse process of the atlas was hypertrophic and displaced laterally toward the styloid process, the transverse process impinged on cranial nerves IX–XII.

Treatment of Collet-Sicard syndrome depends on the underlying cause. There are three reported cases of Jefferson fracture causing Collet-Sicard syndrome [1,14,15]. Two patients with Collet-Sicard syndrome caused by Jefferson fracture were treated with halo traction [1,15]. One patient presented with hoarseness and difficulty in speaking, and 2 weeks of traction, 10 weeks in a halo vest, and 2 weeks in a cervical collar resulted in near resolution of his neurologic symptoms [1]. The other patient with hoarseness and difficulty in swallowing was able to drink water well enough to allow removal of the feeding tube at 8 weeks, but his voice was still abnormal [15]. At 13 months, his voice had normalized, but he still could not sing high notes accurately [15]. Another patient with hoarseness, difficulty in speaking and swallowing, and easy choking was treated with a cervical collar [14]. At 18 months, his only residual symptom was difficulty in speaking [14]. In our patient, however, Collet-Sicard syndrome was due to developmental abnormalities of the craniocervical junction, not due to Jefferson fracture. When neurologic symptoms associated with assimilation of the atlas occur, occipital cervical fusion or decompression is suggested [21]. However, it is not known whether occipital cervical fusion or decompression can treat Collet-Sicard syndrome resulting from assimilation of the atlas.

It is difficult to explain why Collet-Sicard syndrome developed so late in our patient. Although assimilation of the atlas is congenital, most patients do not develop neurologic symptoms until the third or the fourth decade of life [21]. Symptoms progress slowly in the majority of patients, but can develop dramatically in some patients [21]. The degenerative changes of aging might have restricted motion of the lower cervical spine, placing an increased demand on the capsular and ligamentous structures of the atlantoaxial articulation [21]. Furthermore, ligamentous laxity caused by aging might have increased atlantoaxial instability gradually, producing cranial nerve compromise [21]. In addition, young patients' cranial nerves may be more resistant to compression, which later prove harmful to older patients' cranial nerves.

Swallowing difficulty can increase morbidity and mortality in patients with Collet-Sicard syndrome. A water swallowing test is a simple bedside evaluation for dysphagia [22]. However, it has its limitations such as lack of reliability, subjectivity, the possibility of missing patients who are aspirating, and an inability to provide any information regarding the specific mechanism of dysphagia [22]. Therefore, the videofluoroscopic modified barium swallow study has become a standard method [22]. Patients with dysphagia require food of modified consistency [22]. Patients who are unable to orally maintain adequate hydration or nutrition are fed through nasogastric tubes [22]. Dysphagia treatment involves compensatory strategies and rehabilitation strategies [22]. Compensatory strategies are directed at keeping the patient safe with oral intake while the patient undergoes active rehabilitation [22]. Rehabilitation strategies are aimed at changing swallowing physiology and accelerating the recovery process [22]. Because resolution of dysphagia may occur over the weeks and months following damage to cranial nerves, reassessment may be needed within a few weeks of the initial evaluation for dysphagia [1,14,15]. If a second fluoroscopic modified barium swallow study finds no evidence of aspiration, the nasogastric tube can be removed and resumption of a regular diet can be allowed [22].

Conclusions

In suspected Collet-Sicard syndrome, developmental abnormalities of the craniocervical junction should be considered in the differential diagnosis. Patients with known developmental abnormalities of the craniocervical junction who had been treated with conservative measures can develop Collet-Sicard syndrome during follow-up. Surgical therapy with decompression of neural structures might be considered in these patients.

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