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Case Report

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

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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

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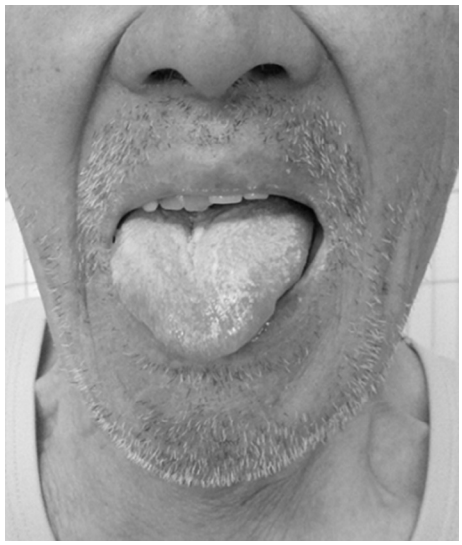

Fig. 1. Photograph of the patient. His tongue deviates to the left.

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

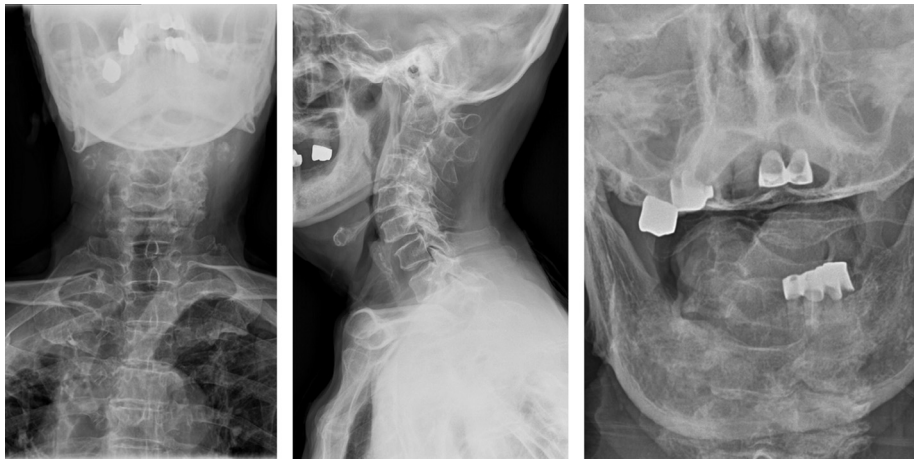

Fig. 2. Cervical spine radiographs. Anteroposterior (Left), lateral (Middle), and open-mouth odontoid (Right) radiographs show an occiput–C1–C3 fusion, hypoplastic dens, and scoliosis.