Collet-Sicard Syndrome Related To Occipital Condyle Fracture. A Case Report

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Abstract:

Collet-Sicard syndrome is a rare clinical manifestation of unilateral palsy of the mixed cranial nerves (from IX to XII). A 37 year-old man was presented, three weeks after a trauma of the left cranio-cervical region, for hoarseness voice, difficulty swallowing, left neck pain and weakness in the left shoulder. The neurological examination uncovered dysfunction on the left side of cranial nerves IX, X, XI, and XII. A cervical computed tomography scan detected a fracture of the left occipital condyle at the skull base. The patient was managed conservatively by immobilization of the cervical spine and orthophonic rehabilitation. Total neurological recovery was noted one year after the injury.

Keyword: Collet-Sicard syndrome; occipital condyle; skull base fracture; cranial nerves palsy

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I. Introduction

Collet-Sicard syndrome (CSS) is a rare clinical manifestation defined as unilateral palsy of the mixed cranial nerves IX, X, XI and XII caused by skull base disease involving the jugular and hypoglossal foramina [1]. Frederic Collet was the first to describe CSS in 1915 during the World War I in a soldier who was victim of a bullet mastoid trauma [2]. In 1917, Jean A. Sicard described five cases similar cases caused by bullet injury [3]. Several causes have been reported including skull base traumatic, vascular, infectious, inflammatory, neoplastic and iatrogenic lesions [4-5]. This report describes a case of palsy of mixed nerves caused by occipital condyle fracture.

II. Case Presentation

We present a case of 37 year-old man received three weeks after a trauma of the left cranio-cervical region secondary to a stick hit. He developed gradually a hoarseness voice and difficulty swallowing for both solids and liquids. He also presented a left neck pain exacerbated by movement and weakness in the left shoulder.

Neurological examination revealed whispering speech, liquid regurgitation, absent gag reflex, left vocal cord paralysis, asymmetric elevation of the soft palate through the right side, tongue deviation and paresis of lateral motion on the left side without fasciculations. We noted atrophy and palsy of the sternocleidomastoid and trapezius from the left side. No mass was felt in the neck. The rest of the neurological evaluation was normal.

Biological analyzes were normal. A cervical computed tomography scan (CT) revealed a left occipital condyle fracture at the base of the skull with no cerebral involvement (**Figure 1**). Magnetic resonance imaging (MRI) of the brainstem, cervical cord and plexus revealed no abnormalities.

We managed our patient conservatively by immobilization of the neck with a collar. A nasogastric tube was not necessary. The patient was seen regularly after discharge at follow-up as an outpatient. Patient had benefited from orthophonic and motor rehabilitation. Three months functional outcome was marked by the improvement of the hoarseness voice, swallowing difficulties and the whispering speech. One year later, we noted a complete recovery of the neurological symptoms.

Figure no 1: Displays axial (A) and left sagittal (B) reconstructed computed tomography scan images using a bone window of the neck, revealing a comminuted fracture of the occipital condyle at the skull base (arrow).



III. Discussion

The IX, X, and XI cranial nerves arise from the collateral groove located on the posterior bulb. The XII nerve originates vertically within the medulla and emerges from the midbrain in 10 to 15 sheaths, forming a nerve root. Its exit from the cranial vault occurs through the hypoglossal canal near the occipital condyle [6]. Affections of the mixed nerves at the base of the skull define clinically Vernet syndrome or Jugular Foramen syndrome. When associated with XII nerve palsy, it define Condylo-jugular syndrome, also known as CSS [7]. The most common cause of CSS is tumor metastasis of skull base, vascular disease, inflammatory processes, trauma and iatrogenic complications [5]. Skull base fracture had been reported as a rare cause of CSS [8]. Occipital condyle, Jefferson and styloid process fractures are documented as causes of post traumatic CSS [4-6-9].

Bell first documented Occipital Condyle Fracture (OCF) in 1817 during an autopsy case [10]. OCF represents a distinct anatomical and clinical entity due to its unique characteristics. The occipital condyles are closely associated with the hypoglossal canal and the jugular foramen, housing cranial nerves IX, X, and XI. Moreover, the occipital condyles hold significant anatomical connections to the brainstem and vascular structures [11]. This close adjacency of the occipital condyle to the jugular foramen elucidates the involvement of the lower four cranial nerves in cases of condyle fracture [12].

The pathogenesis of neurologic deficit was ischemia induced or mechanical lesions [13]. Furthermore, a mechanism of compression and stretching of the nerves has been suggested [14]. Indeed, there have been reports highlighting the potential involvement of displaced bony fragments in compressing nerves, particularly in cases where the fracture extends into the posterior jugular foramen or the anterior condylar canal [15]. All these situations lead to increase inflammation and oedema surrounding the nerve compression.

The diagnosis of CSS can be elaborated through an association of clinical history, physical examination, and analysis of lesions provided by brain imaging [5]. Clinical manifestations vary according to the severity of nerve involvement. Symptoms may include phonation disorders and aspirations such as tracheal aspiration and nasal regurgitation of liquids. Upon examination, motor paralysis of various structures on the ipsilateral side may be observed, including the vocal cord, soft palate (indicated by Curtain's sign), superior pharyngeal constrictor, sternocleidomastoid, and trapezius. Sensory deficits may result in ipsilateral paraesthesia affecting the larynx, pharynx, posterior part of the tongue, and soft palate. Hypoglossal nerve palsy can lead to atrophy of the ipsilateral hemitongue [6]. The introduction of MRI, allowing for coronal and sagittal sections of the skull base and three-dimensional reconstruction, has significantly improved the precision of OCF diagnosis. CT scan with three-dimensional reconstruction is considered the most specific investigative tool for detecting OCF [16].

The management of CSS involves various strategies which include conservative treatment, medical management or surgical treatment. The primary goal of managing swallowing disorders initially is to prevent pulmonary aspiration. Therefore, normal oral intake should be avoided, and enteral feeding can be provided through a nasogastric tube. Typically, this method of feeding is temporary as normal swallowing function often recovers within days to weeks [6]. Orthopedic treatment may involve immobilizing the cervical spine using a collar [14]. Surgical intervention consists on nerve decompression and facture stabilization [17]. This surgical management could have some risks to the patient [18].

For our patient, we opted for a medical treatment. In the majority of posttraumatic CSS cases, neurological recovery is gradual with partial improvement of symptoms. It's noteworthy that residual

neurological deficits may persist for an extended period [11]. Also, it was reported that immediate deficits have a lower rate of recovery than secondary deficits. A total recovery in patients presenting CSS secondary to skull base fractures was reported to be 23% in all cases [6]. In our case, slow total neurological recovery was noted.

IV. Conclusion

The case report summarizes the anatomical associations between the skull base and the mixed cranial nerves. When assessing skull base fractures, the preferred diagnostic modality is a CT scan using serial coronal and sagittal sections. Pulmonary aspirations should be prevented as soon as possible. The conservative management is preferred than surgery. Usually, patients with post traumatic CSS had a poor prognosis. However, a total recovery is possible as we demonstrated in our case.

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