Collet-Sicard syndrome: clinical and radiological aspect

Abstract

Introduction: Vernet, Collet-Sicard, and Villaret syndromes are rare; they affect the pathways of certain cranial nerves and may result in impaired tongue movement. The causes are diverse. Patients with this disorder may experience changes in tongue activity and function, dysphonia, dysarthria, and difficulty swallowing. The purpose of this article is to provide a case report of Collet-Sicard syndrome.

Case report: We report the case of a 72-year-old woman who presented with left Collet-Sicard syndrome. The complete work included a scannographic study and magnetic resonance imaging, and allowed to show the suspected diagnosis was glomus jugulare tumour or infectious processes.

Discussion: Collet-Sicard syndrome is a rare disorder, defined by unilateral paralysis of the last four cranial nerves (IX to XII). It differs from Villaret syndrome in that there is no associated sympathetic involvement. Classical etiologies of Collet-Sicard syndrome include skull base tumors, trauma and vascular damage**.**

Conclusion: Sicard collar syndrome is a rare syndrome leading to discussion of several diagnoses and management is difficult. The Dentist must be familiarized with the functional and anatomic changes in the tongue, and is the professional most indicated for identifying diseases that affect the glossopharyngeal, accessory and hypoglossal nerves

Introduction

Collet-Sicard syndrome is a rare condition, defined by unilateral and simultaneous paralysis of the last four cranial nerves in the posterior condylo -torn intersection, associating IXth, Xth, XIth and XIIth cranial pairs. The usual causes are tumors of the parotid, carotid glomus, or metastatic tumors. It is distinguished from Villaret syndrome by the absence of sympathetic cervical involvement.

Clinical case

A 72-year-old patient with no particular pathological history whose reason for consultation was the onset of otorrhea

Chronic with right otalgia rebellious to medical treatment the evolution was marked by the installation of dysphonia and dysphagia to liquids with right laterocervical pain the clinical ENT and neurological examination revealed a beating mass on otoscopy right retro tympanic, right lingual deviation, typical unilateral paralysis of the soft palate with deviation of the uvula (curtain sign), hypoesthesia to touch and prick of the posterior part of the tongue and, above all, taste disorders: ageusia of the posterior third of the tongue, especially marked for bitter substances, we also noted a lowering of the shoulder stump and difficulty in raising it in connection with paralysis of the trapezius,

with difficulty in the rotational movement of the head, in connection with paralysis of the sternocleidomastoid, and paralysis of the cord of the cord right voice.

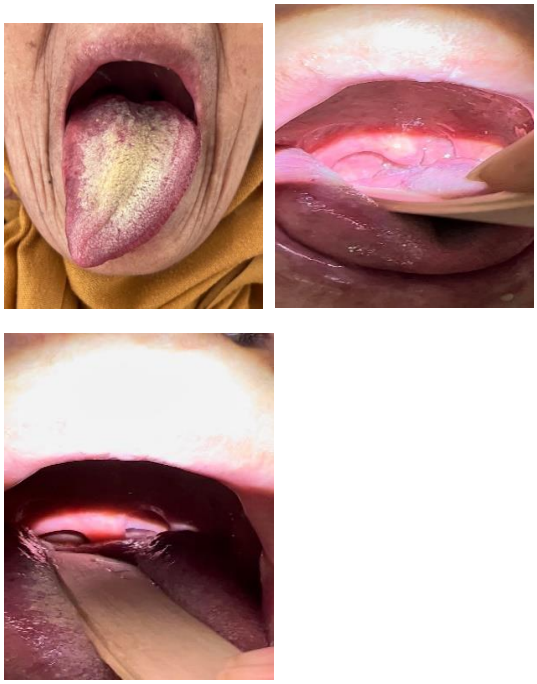

Fig 1 images showing right lingual deviation and velo-palatal paralysis with uvula deviation

VIDEO PARALYSIE VELO-PALATINE.mp4

otoscopie masse battante rétro tympanique droite.mp4

The diagnosis of Collet-Sicard syndrome was made due to this paralysis of the last four right cranial nerves without associated sympathetic involvement.

A cervical and CT scan of the temporal bone was requested showing the presence of total filling of the tympanic cavity with material of density similar to that of the soft tissues with discreet lysis of the long process of the incus and lysis of the wall. lateral side of the facial canal at the level of its tympanic portion with filling of the mastoid cells and eroded appearance of the wall of the cubicle appearance which may be related either to a paraganglioma of the right ear or to reactive otitis with the presence of posterolateral parietal thickening right nasopharyngeal encompassing the right internal carotid and reducing its caliber

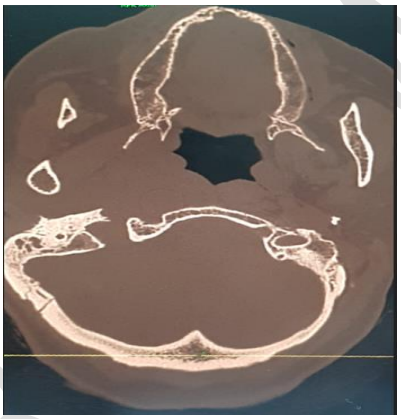

Fig 2 CT image showing the enlargement and filling of the jugular foramen and the Lacerum foramen

On magnetic resonance imaging, presence of a lesional process centered on the right carotid space infiltrating and fusing along it from the carotid bifurcation at the base of the skull with reduction in the caliber of the jugulocarotid axis this process is poorly limited tissue signal in t1 and t2 enhances intensely after injection of the contrast product pushing back the right posterolateral walls of the cavum with normal thickening of its mucosa infiltrating the prevertebral space and filling the lacerum and jugular foramen which are widened with the beginning of invasion of the associated right cavernous sinus to an abnormal bone signal with contrast enhancement of the occipital condyle of the Clivus and the body of the Sphenoid without associated suspicious cervical adenopathy.The suspected diagnosis

was glomus jugulare tumour or infectious processes.

2 biopsies of the cavum under local anesthesia were done with anapath and no signs of specificity or malignancy

We requested a neurosurgical evaluation in order to determine the best treatment approach. Given the patient's advanced age, surgical resection was ruled out. Since we opted for treatment with stereotactic radiosurgery, no anatomical pathology diagnosis of the lesion is available. After 5 years of follow-up, CSS has not yet resolved.

Discussion

The paralysis of the last cranial nerves can be grouped according to three modalities: (1) Either the damage concerns only the nerves which pass through the posterior torn hole (IX, X, XI), producing the syndrome of the posterior torn hole described by Vernet. (2) Or else to this damage of the IX, X, XI pairs is added that of the hypoglossal. It was in September 1914 that Collet observed in a war wounded a syndrome characterized by hemiplegia of the soft palate, larynx, pharynx and tongue, and which he designated under the name of total syndrome of the last four nerves; from this first observation he insisted on the importance of dysphagia, particularly marked for swallowing solids. Sicard and Bollack, in 1912, had described under the name "lingual laryngo -pharyngeal and cleido-trapezial hemiplegia with tachycardia" a very similar syndrome with marked acceleration of the pulse, but where the pharyngeal symptoms were less detailed. This syndrome still bears the name of Collet and Sicard or that of " posterior condylo -torn crossroads syndrome" because of the torn posterior hole through which the IX, X, XI pairs pass, and the condylar hole which gives passage to the XII. (3) Finally, this paralysis of the last four cranial nerves can be superimposed on that of the upper cervical sympathetic nerves, as Villaret showed by providing, in 1916

Anatomically, the glossopharyngeal, vagus and accessory nerves emerge from the base of the skull through the jugular foramen and the hypoglossal nerve through a proper orifice. These nerves group together in the retrostyloid space where they rub shoulders with the internal carotid artery, the jugular vein, the sympathetic and lymphatic nodes (Walker et al. 2003).

This syndrome usually develops gradually and its clinical presentation may be complex, which is why late diagnosis is not infrequent. Sometimes it presents in an incomplete form, resulting in other types of syndromes known as jugular foramen syndromes (Table 1). When CSS appears in association with ipsilateral Horner syndrome, this is called Villaret syndrome.

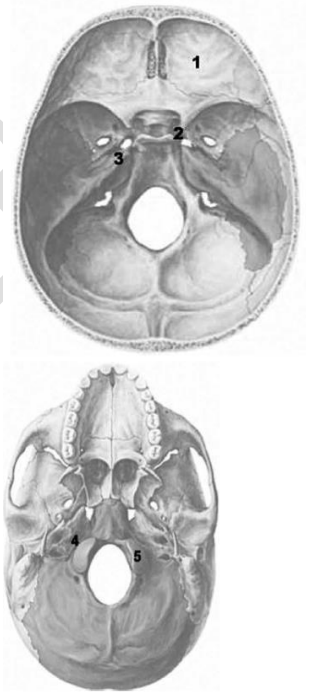

Figure the base of the skull: 1 anterior cranial fossa 2 the parasellar region 3 jugular foramen 4 occipital condyle 5 middle cranial fossa

Syndrome	Affected cranial nerves
Collet-Sicard syndrome	IX, X, XI, XII
Villaret syndrome	IX, X, XI, XII, sympathetic chain (Horner syndrome)
Vernet syndrome	IX, X, XI
Jackson syndrome	X, XI, XII
Schmidt syndrome	X, XI
Tapia syndrome	X, XII

Table 1.Jugular foramen syndromes

Diagnosis of CSS can be based on clinical history, physical examination, and detailed description of the lesions provided by neuroimaging.(4) Occasionally, as in our case, it may be difficult to describe tumours located in the jugular foramen. Gadolinium-enhanced MRI is the technique of choice for determining tumour size and anatomical connections. However, an anatomical pathology study is necessary for a definitive diagnosis. (5)

Causes can be divided into neoplastic (jugular paraganglioma, schwannoma ,metastases) and non-neoplastic (Trauma, osteomyelitis ,Paget disease, vascular disorders). Our patient showed clinical findings compatible with CSS secondary to a primary tumour in the jugular foramen, possibly related to the presence of a glomus jugulare tumour. Glomus tumours, or paragangliomas, are highly vascularised tumours composed of cells that originate from the neural crest during embryonic development. While 90% of the paragangliomas arise in the adrenal glands, only 3% develop in the head and neck. (7) These tumours grow close to the jugular foramen and may extend into the intracranial and extracranial spaces. Surgical resection is the treatment of choice for neurinomas and paragangliomas. However, stereotactic radiosurgery is the main treatment alternative for elderly patients, those in poor clinical condition, or patients presenting residual or recurrent lesions after surgery.(8),Signs com compromised motricity and lingual anatomy must be carefully evaluated by the Dentist. The possibility of compromise of the sensitive and motor nerves that innervate the region must always be investigated by means of complementary exams (tomography, magnetic resonance imaging, rhinoscopy, laryngoscopy) when no local cause is identified in the intraoral evaluation. Precise diagnosis is frequently neglected, and the patient goes without precise diagnosis and ideal treatment for months or years.

A search of the literature on CSS in PubMed identified 51 cases published between 1915 and 2012. Table 2 summarizes the various reasons for CSS. Of all skull base tumor-related processes that may lead to this syndrome, metastasis is the most common cause. We found only three cases of CSS secondary to primary tumors of the skull base: two secondary to jugular body tumors9, 10 and one secondary to hypoglossal nerve schwannoma. 11

Highlights

Describe and study the case of a rare Collet-sicard syndrome ¬ highlight the role of the MRI in the assessment of the extension and the histological study to make the diagnosis ¬ The complete work included a scannographic study and magnetic resonance imaging, and allowed to show the causal compressive lesion in the retroparotid space.

Conclusion

The current approach to Collet-Sicard syndrome is of extreme relevance to the medical community as it is a rare event with few reported cases. Given this, it is important to ask whether the reported syndrome is an underdiagnosed disease or whether it constitutes a truly rare condition. Therefore, investments in research and studies in this area are extremely relevant.

There is no large database on the rare syndrome, therefore, studies on comorbidity are still needed in order to improve the associated symptomatology. It is therefore fundamental to continue research into Collet-Sicard syndrome, including its symptoms, diagnosis, treatment and clinical course,

because it is a rare condition and still little described in the literature.

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