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Collet–Sicard Syndrome Caused by Spontaneous Internal Carotid Artery Dissection: A Case Report and Review of Literature

CASE REPORT
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ABSTRACT

Dissection of the internal carotid artery (ICA) is a serious and acute event that can present with different clinical pictures. In most cases, dissection involves the intracranial portion of the internal carotid artery, and depending on the location and extent of the vascular injury, symptoms should include hemiplegia, visual impairment, ophthalmoplegia, and loss of consciousness. In clinical practice, however, it is important to consider the possibility of ICA dissection in the neck or in the region of the carotid foramen at the base of the skull and its association with atypical symptoms such as acute and progressive mixed cranial nerve deficits in the absence of other neurological signs. In any case, ICA dissection requires a rapid and effective physical examination (neurologist, anesthesiologist, otolaryngologist) and neuroradiological assessment. We report a rare case of Collet–Sicard syndrome due to spontaneous dissection of the extracranial part of the ICA and include a detailed review of the literature.

Keywords: Collet–Sicard syndrome, cranial nerve palsy, internal carotid artery dissection

Introduction

Collet–Sicard syndrome (CSS), characterized by a unilateral deficit of the cranial nerves IX-X-XI-XII, is a rare clinical condition first described in 1914 by Frederic Collet in a World War I soldier after a bullet hit the mastoid region.[1] A few years later, in 1917, Jean Sicard described a case of craniovertebral junction (CVJ) trauma with paralysis of the last 4 cranial nerves.[2] Subsequent anatomical and radiological studies of a case report have shown that the etiopathology of the traumatic event lies in the transmission of traumatic energy through the base of the skull and the CVJ, with occlusion of the jugular foramen by a displaced bony fragment.[3] Collet–Sicard syndrome can also be provoked by vascular diseases, surgery of the posterior fossa and lacerum foramen, or skull base tumors. Damage to the vascularization of the posterior fossa exposes the bulbar region of the brainstem to ischaemia.[4] The vertebrobasilar system and its collateral branches to the brainstem and cerebellum may be the primary site of pathology, as in thrombotic and aneurysmal events, or secondary to systemic pathologies such as Wegener's granulomatosis, dysmetabolic, and infectious diseases.[5-6] Dissection of the internal carotid artery (ICA) may be responsible for lower cranial nerve deficits in 3% of cases. The incidence of ICA dissection ranges from 2.6 to 2.9 per 100 000 cases, with an average age between 40 and 45 years and no male/female preponderance.[7] Dissections of the extracranial portion of the ICA are much more common than intracranial dissections, accounting for more than 90% of all carotid dissections. The extracranial dissection usually starts close to the C2-C3 vertebral level and continues for varying lengths, stopping at the entrance to the carotid canal in most cases. The aim of this paper is to report an interesting new case of lower cranial nerve palsy following spontaneous ICA dissection and to review the literature (Table 1).

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Table 1. List of Collet–Sicard Syndrome Cases Caused by Spontaneous Internal Carotid Artery Dissection Reported in the Literature

Authors (years)	No. of cases	Diagnosis	Management	Outcome
Mokri et al (1992)	3	Angiography	Conservative/Anticoagulants	2 patients improved 1 patient recovered
Rees et al (1997)	1	MRI/Angiography	NA	Recovered
Heckmann et al (2000)	1	Ecodoppler/Angiography	Conservative/Anticoagulants	Improved
Mohr et al (2005)	1	NA	Conservative/Anticoagulants	Improved

Case Presentation

B.C., a 72-year-old man, was admitted to the hospital after complaining of dysphonia, dysphagia, and a left-sided headache refractory to common analgesics. Examination revealed left tongue deviation and soft hemipalsy. Fiberoptic evaluation also revealed left vocal cord paralysis. There was also a contractile deficit in the left sternocleidomastoid and trapezius muscles. There were no other neurological symptoms, in particular no other cranial nerve deficits, visual acuity, or oculomotion deficits. The brain CT was normal and showed no focal lesions or signs of stroke. An MRI of the skull base and neck showed a dissection of the left ICA at the level of the carotid foramen. In particular, T1-weighted spin echo images acquired in the axial plane showed an eccentric hyperintense signal surrounding the left ICA in the distal part of the petrosal region with a subacute intramural hematoma (Figure 1). In addition, on coronal T2-weighted images, there was a clear cord-shaped signal defect along the ICA at the level of the first 2 vertebrae (Figure 2). We also report one of the most important radiological images of the angiographic MRI sequences, showing the extent of the left carotid dissection (Figure 3). Blood tests, including coagulation factors, lupus-like anticoagulant, antiphospholipid antibodies, and homocysteine, were normal. The patient was treated with oral anticoagulants (INR range: 2-3) and gradually improved until complete clinical recovery.

Ethics

The consensus statement was prepared according to the CARE guidelines. Written informed consent was obtained from the patient for the collection of data for scientific purposes.

MAIN POINTS

- *Collet–Sicard syndrome (CSS) is characterized by paralysis of the last 4 cranial nerves. When caused by vascular phenomena, the presence of symptoms such as headaches, visuomotor deficits, and facial dysesthesia is likely to depend on secondary peripheral microembolic phenomena.*
- *Vascular causes (ICA involvement) account for about 3%-5% of all CSS cases. Most cases are due to craniocerebral trauma, oncological causes (primary tumors or metastases of the jugular foramen region), infectious (skull base osteomyelitis), or iatrogenic (e.g., infratemporal type A approaches, petro-occipital approaches).*
- *Irrespective of the determining cause, which may require endovascular, fibrinolytic, or surgical treatment (decompression, trauma, etc.), the patient affected by CSS requires timely logopedic management to optimize the recovery margins of nerve function and limit the risk of complications such as pneumonia, ab ingestis, and dysphagia.*

Discussion of Pathogenic Mechanisms and Review of Literature

Collet–Sicard syndrome is a rare condition characterized by unilateral paralysis of the last 4 cranial nerves, described by the otolaryngologist Frederic Collet and the neurologist Jean Sicard. It is distinguished from Villaret's syndrome by the absence of sympathetic manifestations and from Vernet's syndrome, in which the XII cranial nerve is not affected. Diseases affecting the blood vessels, such as diabetes or connective tissue disorders, can cause cranial nerve palsies.[8-9] Many reports of ICA dissection associated with hypoglossal palsy or lower cranial nerve palsy have been attributed to mechanical compression and stretching of the nerves below the jugular foramen by the dilated or aneurysmal ICA involved in the dissection. However, although the so-called mechanical theory is important in the etiopathogenesis of lower cranial nerve palsy, it does not explain the clinical cases in which the symptomatology of nerve palsy deficiency occurs

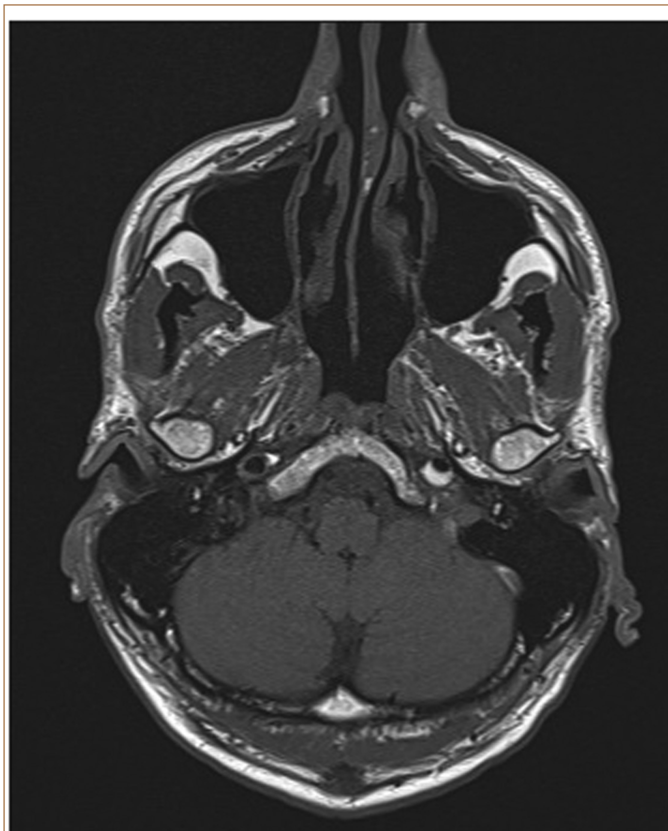

Figure 1. The non-contrast MRI sequence is T1-weighted and shows a circumferential dissection of the left ICA wall.

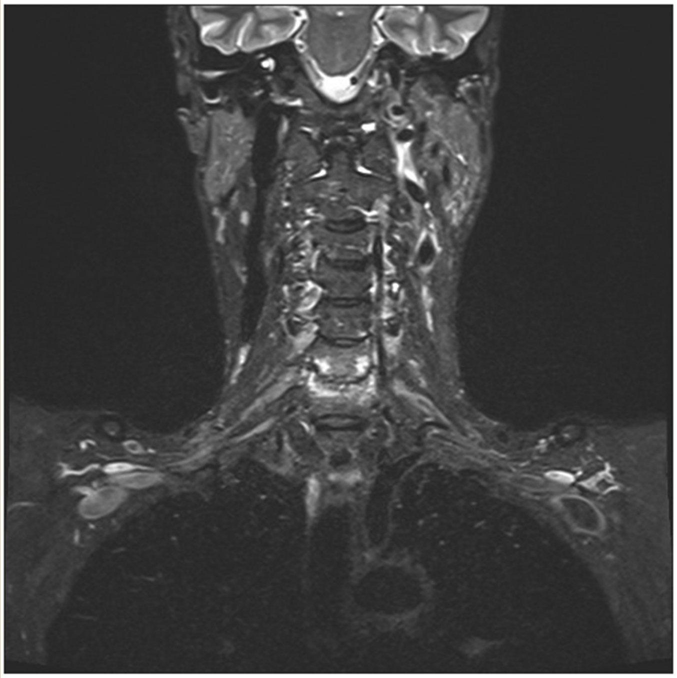

Figure 2. In the coronal T2-weighted sequence, the image shows the dissection of the left ICA in its cranio-caudal extension. The false lumen is visible as hyperintense, following a tortuous course of the artery.

in the absence of aneurysmal dilatation of the ICA. Similarly, the theory of mechanical compression by aneurysmal dilatation cannot explain cases in which lower cranial nerve palsy is associated with hemifacial paresthesia due to trigeminal deficiency or oculomotor deficit due to oculomotor involvement. To explain these cases, a more plausible mechanism for cranial nerve palsy in ICA dissection is impairment of the blood supply to the cranial nerves. This may be mechanical, embolic, or hemodynamic. An anatomical autopsy study described a very small artery that sometimes leaves the ICA near the base of the skull.[10] If this small artery contributes to the vascular supply of nearby cranial nerves, its impairment may cause segmental ischemic damage to these nerves. The main carotid vascular systems supplying the cranial nerves are the inferolateral trunk, the middle meningeal system, and the ascending pharyngeal system. In particular, the pharyngeal system supplies cranial nerves IX to XII.[11] Several variations in the collateral arterial supply to the cranial nerves may occur; for example, an ascending pharyngeal artery may arise anomalously from the cervical ICA and supply the lower cranial nerves. The radiological diagnosis of ischemic occlusions of these small arterial trunks is difficult on MR angiographic sequences but can be demonstrated with selective angiography. We conclude that cranial nerve palsies are seen in 10%-20% of patients with extracranial ICA dissection.[12] Although compression or stretching of the cranial nerves by the dilated artery may explain some of these cranial nerve palsies, the majority are likely to result from a compromise of the nutrient arterial supply to the relevant nerve. This impairment may be caused by distal embolization or by the anomalous origin of the nutrient vessel and its compromise following dissection. The abovementioned pathogenetic mechanisms of the neural deficit and the possible consequences for the intracranial microcirculation may explain the clinical heterogeneity of the cases of cranial nerve palsy during carotid dissection reported in the literature (Table 2). Although acute and chronic vascular pathologies are important causes of CSS, it is important to consider the wealth of literature on traumatic, oncological, and infectious causes. In fact, it is well known that cranio-vertebral trauma, especially in the occipital and cervical regions, can cause medullary and intracranial injuries. In this context, the Jefferson fracture has been associated with the development of CSS. The pathophysiology of lower nerve deficits results from direct trauma (with nerve laceration) caused by free bone fragments or by crushing/compression of the nerve following displacement of the vertebral-occipital joint.[13] The neurovascular structures of the skull base and CVJ may also be affected by ischemia or infiltration caused by primary or secondary neoplastic lesions. The most common oncological lesions are metastases from various sites. A recent study, evaluating the incidence and histopathological characteristics of temporal metastases in cancer patients showed a greater tendency for skull base involvement by lesions originating from the breast, lung, and prostate. The common feature of all types of metastases was osteolytic damage to the region of the jugular foramen.[14] One of the first case reports of breast cancer metastasis and CSS dates back to 1980. Although traumatic, oncological, and vascular causes are among the most serious in terms of prognosis, it is worth mentioning that there are infectious causes that can damage veins and nerves that cross the skull base. This is the case of severe temporal osteomyelitis, as reported by Sibai et al They reported a case of a patient who developed osteitis of the lateral skull base as a complication of otitis media.[15] Finally, it is important to specify how another potential cause of CSS is represented by the iatrogenic cause. This is the case in patients undergoing jugular foramen and lateral skull base surgery (e.g., infratemporal approaches type A, petro-occipital approaches to the posterior fossa).

Figure 3. The coronal angio-MRI shows the width of the false lumen in the left ICA and shows an increase in the maximum diameter of the artery.

Table 2. List of Lower Cranial Nerve Palsy Cases Caused by Vascular Diseases Reported in the Literature

Authors (years)	Cranial Nerve Involvement	Cerebral Ischemia	Headache	Sympathetic Palsy
Lane et al (1980)	IX-XII	None	Yes	Yes
Bradac et al (1981)	IX through XII (various involvement in 2 cases)	None	Yes	Yes
Havelius et al (1982)	IX through XII	None	None	None
Goodman et al (1983)	XII	Yes	Yes	Yes
Goldberg et al (1986)	IX through XII	None	Yes	Yes
Lieschke et al (1988)	X-XII	None	Yes	None
Waespe et al (1988)	IX through XII	None	Yes	None
Bradac et al (1989)	X-XII (variously involved in 3 cases)	Yes (variously involved in 3 cases)	Yes (variously involved in 3 cases)	Yes (variously involved in 3 cases)
Dal Pozzo et al (1989)	IX through XII (various involvement in 3 cases)	None	Yes (variously involved in 3 cases)	Yes (variously involved in 3 cases)
Panisset et al (1990)	IX through XII (various involvement in 3 cases)	None	Yes	None
Vighetto et al (1990)	XII	None	Yes	Yes
Schievink et al (1990)	IX-X-XII	None	Yes	Yes
Hess et al (1990)	IX through XII	Yes	Yes	None
Mokri et al (1992)	IX through XII (various involvement in 8 cases)	Yes	Yes	Yes (one case)
Sturzenegger et al (1993)	XII	None	Yes	Yes (one case)
Rees et al (1997)	IX through XII	None	None	None
Heckmann et al (2000)	NA	NA	NA	NA
Mohr et al (2005)	IX through XII	None	None	None
Nguyen et al (2013)	XII	None	None	None
English et al (2018)	IX-X-XII	Yes	Yes	None
Saliou et al (2018)	IX through XII	Yes	None	None
Peters et al (2018)	VII-VIII	None	None	None
Mes et al (2018)	XII	None	None	None

Collet–Sicard Syndrome: Therapeutic Options

The wide etiological variability of Collet–Sicard syndrome requires different therapeutic options. Thromboembolic problems or segmental dissection of the ICA can be managed conservatively with anticoagulation or thrombolysis. However, in cases where the thromboembolic or dissection picture is more extensive, endovascular interventional procedures may be required. Carotid stenting can be lifesaving but has the risk of complications such as wall rupture. The literature is very heterogeneous regarding the management of post-traumatic CSS; many authors emphasize the importance of surgical approaches aimed at stabilizing the fracture lines of the cervical spine and CVJ. The management of oncological causes must be referred to a multidisciplinary tumor board. Finally, infectious causes of CSS require microbiological characterization in order to initiate specific antibiotic therapies; they may sometimes require extensive surgical treatment. In any case, it is essential that the patient with a deficit of the last 4 cranial nerves receives specialist and speech therapy support in order to achieve the best possible functional recovery.

Discussion

Collet–Sicard syndrome is a rare syndrome characterized by paralysis of the last 4 ipsilateral cranial nerves. Although the causes are varied, the physiopathology of the clinical picture is often related to primary or secondary vascular neuropathy. Acute dissection of the ICA is, in most cases, confined to the extracranial portion and may not be associated with focal neurological symptoms. Early diagnosis of this condition may limit neurological damage and ensure partial or complete recovery of nerve function.

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