Carotid fibromuscular dysplasia and paresis of lower cranial nerves (Collet-Sicard syndrome)

Case report

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A patient suffered the acute onset of unilateral paresis of the ninth through 12th cranial nerves (Collet-Sicard syndrome). Ipsilateral retrograde jugular phlebography and carotid angiography revealed irregular aneurysmal changes of the internal carotid artery at the base of the skull, causing compression of the internal jugular vein below the jugular foramen. This finding is discussed in relation to the clinical symptoms and signs, and possible mechanisms are examined. Family history as well as the clinical and roentgenological findings were compatible with a diagnosis of fibromuscular dysplasia.

KEY WORDS – carotid aneurysm • fibromuscular dysplasia • glossopharyngeal nerve • vagus nerve • accessory nerve • hypoglossal nerve • jugular foramen syndrome • Collet-Sicard syndrome

Unilateral lesions of the lower cranial nerves (ninth, 10th, 11th, and 12th: the Collet-Sicard syndrome) are rare. This variant of the jugular foramen syndrome was described in the victims of World War I by Collet and Sicard, and penetrating injury has remained a rather frequent cause. However, the syndrome may also be due to various neoplasms, and inflammatory and vascular lesions. Among vascular disorders, an aneurysm of the internal carotid artery (ICA) at the base of the skull has rarely been reported as the presumptive cause.

In the case reported here, a unilateral Collet-Sicard syndrome was believed to be caused by an extracranial carotid aneurysm. The underlying vascular disorder was considered to be fibromuscular dysplasia, which has only recently been associated with lesions of the lower cranial nerves.

Case Report

This 44-year-old man had previously been in good health except for arterial hypertension for a couple of years, which had not been treated. Family history was significant with respect to the incidence of arterial hypertension and vascular disorders. Most of his father's relatives had died from cardiocerebral vascular complications, including rupture of an intracranial aneurysm.

On July 20, 1980, the patient complained of a sore throat, hoarseness, slurred speech, and difficulty in swallowing. Taste sense was impaired and he noted several mucosal blisters on the right anterior pillar of the fauces. He was afebrile and otherwise asymptomatic. After a couple of days, he was admitted to his local hospital. Examination at that time revealed unilateral involvement of the ninth through 12th cranial nerves on the right side. Otherwise, neurological examination was normal. The cerebrospinal fluid was acellular, and had a protein content of 0.4 gm/liter. Routine blood laboratory tests were normal, as were radiological examinations of the skull (including tomography of the base of the skull), heart, lungs, and soft tissues of the neck. The patient improved somewhat. Within 1 week of the onset of symptoms, his taste sense had normalized, and dysphagia was less severe. As the symptoms persisted, he was referred to our Department of Neurology for further evaluation.

On admission on August 19, 1980, general examination was normal except for an arterial blood pressure of 190/115 mm Hg. The electrocardiogram was pathological, with tall R-waves and slightly depressed
FIG. 1.  *Left:* Right jugular phlebography, lateral view, showing marked stricture of the right internal jugular vein at the base of the skull (short arrows). The left internal jugular vein appears normal (curved arrow).  *Center:* Right internal carotid angiography, lateral view, showing irregularities of the distal extracranial portion of the right internal carotid artery, with regions of aneurysmal dilatation (arrow).  *Right:* Left internal carotid angiography, anteroposterior view, showing similar but less pronounced changes on the left side (curved arrow).

ST segments in the precordial leads. No cervical bruits were heard, nor were any cervical lumps palpated. The neurological examination disclosed right-sided pareses of the ninth through 12th cranial nerves. There was no ipsilateral Horner’s syndrome. An electromyogram confirmed the unilateral involvement.

An extensive laboratory work-up, including repeated urinary sampling for analysis of adrenaline and noradrenaline, was unrevealing. There were no significant increases in the antibody titers against herpes simplex, varicella, and Coxsackie B 1-6 viruses. The patient was started on a course of metoprolol (Seloken) because of the arterial hypertension.

Right-sided retrograde jugular phlebography (Fig. 1 left), performed to exclude a possible chemodectoma, demonstrated that the internal jugular vein close to the base of the skull was displaced anteriorly, pursuing an arched course with the concavity directed posteriorly. Within this area, there was also marked compression of the vein. However, contrast medium passed through the transverse sinus to a normal left internal jugular vein. Repeat jugular phlebography 1 week later, with a more proximal catheter position, confirmed these findings.

A CT study of the neck and skull base before and after intravenous contrast-medium administration was normal. Right-sided carotid angiography (Fig. 1 center) demonstrated marked pathological changes in the distal extracranial portion of the ICA, with aneurysmal dilatation. Left-sided carotid angiography (Fig. 1 right) disclosed similar, although less marked, changes. Right-sided external carotid angiography was normal. Arch angiography revealed no further abnormalities in the brachiocephalic vessels, whereas abdominal aortography (Fig. 2 left) demonstrated bilateral luminal irregularities in the main renal artery, with aneurysm formation at its bifurcation. Luminal irregularities were also observed in the major intrarenal arteries on both sides, and in the upper part of the left kidney there was an infarct (Fig. 2 right). The angiographic findings were interpreted as compatible with fibromuscular dysplasia.

Over the following year, the patient has continued to improve, although slight dysarthria and dysphagia still persist. No further neurological manifestations have appeared, and thus the left carotid aneurysmal dilatation has remained asymptomatic.

**Discussion**

Aneurysms of the cervical portion of the ICA constitute about 4% of all aneurysms. Considering the topography of the nerves and vessels at the skull base, it is evident that a major carotid aneurysm may displace and stretch the exiting lower cranial nerves and thereby cause corresponding neurological defects. Yet, a survey of the literature indicates that aneurysm is an extremely rare cause of any of the jugular
foramen syndromes. In a thorough review of carotid aneurysms in 1926, Winslow found only five published cases (out of a total of 42) of “spontaneous” aneurysms in patients who exhibited lesions of one or more of the lower cranial nerves. One of these patients, originally reported by Cannuyt, was similar to ours, and suffered from pareses of the ninth through 12th cranial nerves secondary to an ipsilateral carotid aneurysm. Another patient with identical neurological deficits and a carotid fusiform aneurysmal dilatation has been reported by Svien, et al., who reviewed the causes of jugular foramen and allied syndromes in 29 patients.

As a carotid aneurysm at the skull base may remain neurologically asymptomatic, factors other than the size of the aneurysm may sometimes be decisive of whether or not extracranial neurological complications will occur. What are these possible mechanisms and how do they apply to the present case?

Although the temporal profile of the developing symptoms and signs do not necessarily reflect the immediate pathogenesis, it usually provides a sound clinical basis for speculation. Sometimes the neurological manifestations evolve slowly and progressively, a pattern compatible with an expanding mass (that is, the aneurysm) compressing the surrounding tissues. However, in the few published case reports and in our patient, the onset of symptoms has been acute, or subacute, evolving over a couple of days. Such a course may indicate hemorrhage of a dissecting aneurysm, which dislocates and compresses the adjacent nerves. The presence of ipsilateral local or referred pain and/or a Horner’s syndrome is further circumstantial evidence in favor of such a mechanism which is known to complicate fibromuscular dysplasia.

The acute onset of dysfunction of the lower cranial nerves, particularly in the absence of severe pain and with intact ocular sympathetic fibers, may tentatively indicate a slightly different mechanism. In an unpublished study of autopsy material, the two first authors have shown that a very minor artery sometimes leaves the ICA close to the skull base. If this minor artery contributes to the vascular supply of the nearby cranial nerves, an occlusion, either by vascular dissection within the aneurysmal sac or by thrombosis, may cause segmental infarctions in these peripheral nerves.

Another pathogenic possibility to be considered is a viral infection triggered by the traumatic impact of the aneurysm on the cranial nerves. It is well known that a compressive nerve lesion may activate an intra-neuronal varicella virus and cause herpes zoster. Furthermore, surgery on the Gasserian ganglion may provoke facial herpetic labial blisters have been observed in a patient with an aneurysm of the carotid artery arising in the carotid canal and extending to the Gasserian ganglion, causing trigeminal nerve injury. Our patient, like that of Farrell and Ellenberger, initially complained of a sore throat, and reported blisters on the anterior pillar of the fauces on the same side as the subsequent paresis of the cranial nerves. Considering their location and the concomitant change in taste sense, a lesion of the geniculate ganglion or the glossopharyngeal nerve seems probable, particularly of the latter since the facial nerve remained intact. However, an infectious pathogenesis is at least partly discounted by the normal routine laboratory findings in the blood and cerebrospinal fluid during the early stage of the disorder, and serological tests did not provide support for viral infection as the immediate cause.

The natural course with subsequent improvement seen in our case and those of Farrell and Ellenberger and Svien, et al., may be compatible with either of
Carotid aneurysm and Collet-Sicard syndrome

the postulated mechanisms. Neither the neuroradiological evaluation nor the laboratory workup provided definite evidence favoring either etiology.

The combination of bilateral carotid aneurysm dilatations and renal vascular abnormalities found in our patient favors a diagnosis of probable fibromuscular dysplasia. The conspicuous family history with respect to vascular disorders indicated the possibility of familial fibromuscular dysplasia. Only recently has this disease been associated with extracranial carotid aneurysms, causing glossopharyngeal and hypoglossal nerve palsies. Consequently, fibromuscular dysplasia should be considered in the differential diagnosis of jugular foramen syndromes.

References

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