Tenth and Twelfth Nerve Palsies in a Patient with Internal Carotid Artery Dissection Mistaken for Cervical Mass Lesion

Christoph Arnoldner, M.D.,¹ Dominik Riss, M.D.,¹ Jens Wagenblast, M.D.,² Veronika Starlinger, M.D.,¹ and Jafar-Sasan Hamzavi, M.D.¹

ABSTRACT

Among the multiple causes for cranial nerve palsies, internal carotid artery dissection is rather uncommon. Patients usually present with unilateral head pain, Horner's syndrome, and signs of cerebral ischemia. We present the case of a 52-year-old male patient, who showed isolated palsies of the tenth and twelfth nerve without any other symptoms. Magnetic resonance imaging (T1) depicted a hyperintense lesion surrounding the internal carotid artery, which was mistaken for a cervical mass, and the patient underwent unnecessary surgical exploration of the neck. Angiography performed afterward could reveal the dissection of the internal carotid artery. This case shows that even in cases with mild and atypical symptoms, internal carotid artery dissection has always to be ruled out in lower cranial nerve palsies.

KEYWORDS: Carotid artery, dissection, cranial nerve, palsy, radiology

Among a variety of causes for multiple cranial nerve palsies, such as cerebral ischemia, diabetic nerve ischemia, meningitis, subarachnoid hemorrhage, malignant tumors of the skull base, neck, or upper mediastinum, and surgery of the thyroid, spontaneous internal carotid artery dissection (ICAD) is rather uncommon.

The annual incidence of spontaneous carotid artery dissection ranges from 2.5 to 3 per 100,000^{1,2} with a distinct peak in the fifth decade of life.³ In younger patients, ICAD is an important cause of stroke, accounting for ~20% of such cases.⁴

The classical symptom triad of ICAD includes unilateral head, face, or neck pain, Horner's syndrome, and delayed focal cerebral or retinal ischemic symptoms. Nerve palsies can either appear isolated or in various

combinations. In a series of 190 patients with ICAD, Mokri et al⁵ found a syndrome of lower cranial nerve palsies in 5.2% and a syndrome of ocular motor palsies in 2.6% of patients. Cranial nerve palsy without headache is described to occur in only 0.5% of cases.⁵

We describe a rare case of ICAD presenting as isolated cranial nerve (X, XII) palsies, with inconclusive magnetic resonance imaging interpretation, which lead to a distinct delay of correct diagnosis and treatment.

CASE REPORT

A 52-year-old man with no significant medical history presented to the ENT department with dysphagia, hypomotility of the tongue, and hoarseness for the

¹Department of Otorhinolaryngology, Medical University of Vienna, Vienna, Austria; ²Department of Otorhinolaryngology, Johann Wolfgang Goethe University, Frankfurt/Main, Germany.

Address for correspondence and reprint requests: Christoph Arnoldner, M.D., Department AKH-HNO, Medical University of Vienna, Waehringer Guertel 18-20, A-1097 Vienna, Austria (e-mail: christoph.arnoldner@meduniwien.ac.at).

Skull Base 2010;20:301-304. Copyright © 2010 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1 (212) 584-4662.

Received: October 22, 2009. Accepted: January 11, 2010. Published online: March 2, 2010.

DOI: <http://dx.doi.org/10.1055/s-0030-1249246>.

ISSN 1531-5010.

previous 4 days. The detailed interview of the patient revealed a loss of weight during the weeks before presentation, with an acute onset of the complaints mentioned above a few days before. The patient smoked 20 cigarettes per day and negated alcohol consumption. No infectious or traumatic context could be found. Routine ENT examination revealed a deviation of the tongue toward the left with ipsilateral muscular atrophy and paralysis of the left vocal cord. Examination of the other cranial nerves and peripheral nervous system was unremarkable. Routine laboratory investigations revealed no abnormalities. The patient presented to a neurologist, who initiated a magnetic resonance imaging (MRI) of the brain, which was normal. The diagnosis of left X and XII nerve palsy was made.

The patient refused hospitalization, but presented to the emergency ward the next day with subjective deterioration of the symptoms and was then admitted for further examination.

A duplex ultrasound of the carotid arteries revealed no particularities. A computed tomography (CT) of the head and neck showed an asymmetry at the level of the tongue base. Additionally, a contrast medium-enhancing lesion inferior to the jugular foramen, surrounding the left internal carotid artery (ICA) with a caudal extension to the hard palate, was detected. A mass lesion or arteriitis was suspected. An MRI scan (T1-weighted, short tau inversion recovery) showed a lesion surrounding the ICA with moderate to distinct enhancement (Fig. 1). No luminal particularities were

found. The presumed diagnosis of an inflammatory pseudotumor was made.

Therefore, the patient was put on a steroid medication of 250 mg prednisolone daily. On regular ENT examination, no clear amelioration of the palsies could be found. Blood testing for vasculitis (p-ANCA, c-ANCA) and infectious diseases was negative.

An MRI scan after 12 days of steroid treatment revealed no regression of the mass, and therefore a diagnostic biopsy of the suspicious region was indicated. Using a standard cervicotomy approach, the region was examined and a bluish tumor surrounding the ICA was found. During careful dissection of this region, the patient developed an asystolia and needed to receive cardiac massage for a period of 10 seconds. At biopsy, dark-colored blood discharged from the site and consequently no mass could be found any longer. During the postoperative period, no significant amelioration of the symptoms was observed. The histologic examination revealed connective tissue with signs of chronic, fibrous inflammation without signs of neoplasia. Not until 6 weeks after admission, a digital subtraction angiography was performed, which revealed an aneurysma of the left ICA in its most distal cervical part, just inferior to the carotid canal. The long segment dissection of the ICA was not depicted on angiography because of a complete intramural thrombosis of the dissected segment (Fig. 2). CT angiography confirmed the dissection and partial thrombosis of the aneurysma (Figs. 3 and 4). Because of the localization adjacent to the base of the

Figure 1 Magnetic resonance imaging (MRI) scan (T1-weighted, short tau inversion recovery): intramural thrombus appearing as hyperintense signal around left carotid artery. The lesion shows no intraluminal abnormalities and was misdiagnosed as an inflammatory pseudotumor.

Figure 2 Left carotid digital subtraction angiogram: 1 cm aneurysma of the left internal carotid artery (ICA) in its most distal cervical part (arrow), just inferior to the carotid canal. The long segment dissection of the ICA cannot be depicted on angiography due to a complete intramural thrombosis.

Figure 3 Computed tomography showing dissection of the left internal carotid artery (ICA; arrow) with partial preservation of intraluminal blood flow.

skull and the small diameter of the aneurysma (~1 cm), no surgical intervention was considered and the patient was put on an anticoagulant treatment with Phenprocoumon. On checkup 4 weeks later, the patient reported a subjective improvement of the symptoms and showed a clear regression of the palsies on examination.

DISCUSSION

When blood penetrates through an intimal tear and extends within the wall, dissection of the artery results. A subintimal dissection tends to result in narrowing or occlusion of the arterial lumen, whereas a subadventitial

dissection causes aneurysmal dilatation of the artery.⁶ ICAD can either occur spontaneously or following minor trauma or physical effort, such as coughing, vomiting, body building, or chiropractic manipulation.⁷ Congenital tissue defects such as fibromuscular dysplasia, Ehlers-Danlos syndrome type IV, Marfan's syndrome, or autosomal dominant polycystic kidney disease are associated with an increased risk of spontaneous dissections of the carotid and vertebral arteries.⁸ Cranial nerve palsies can be explained by mechanical compression or stretching of the IX to XII nerves, caused by the expanding dissection, or by an interruption of the blood supply to the nerves.⁵ All cranial nerves, except the olfactory, may be affected in different combinations. The hypoglossal nerve was found to be affected most frequently (5%; 5).

Conventional angiography has long been the gold standard in the detection of ICAD, because it shows the arterial lumen and allows extensive characterization of dissection.⁸ The most common finding is a tapered stenotic segment and referred to as string sign. In the last few years, MRI and MR-angiography have largely replaced conventional angiography because of its high resolution and its capability of detecting ICAD, either without associated luminal abnormalities, or with nonspecific luminal occlusion.⁸ In T1-weighted and T2-weighted images, the intramural hemorrhage usually appears as a crescent-shaped hyperintensity that surrounds the narrowed lumen (signal void) and expands the arterial wall.⁹ For three-dimensional time of flight magnetic resonance angiography, a 95% sensitivity and 99% specificity was described, whereas for MRI the values were 84% and 99%, respectively.¹⁰

Duplex sonography is less reliable than angiography and MRI techniques, primarily because the site of dissection is usually far too distal to be seen on sonograms. In addition, sonographic diagnosis can be hindered if the intraluminal blood flow is not altered. Nevertheless, ultrasound can be useful in the initial assessment of patients who are thought to have an ICAD, because an abnormal pattern of flow is identified in >90% of patients.¹¹ Combining ultrasonography with transcranial Doppler can improve the sensitivity of the examination, especially for locations near the base of the skull.¹¹ Contrast-enhanced CT also shows typical abnormalities and is advantageous due to its availability.

To prevent thromboembolic complications, anticoagulation for 3 to 6 months is recommended, although no randomized trials have been undertaken until today to prove the validity of this treatment. If complete recanalization is demonstrated on follow-up MRI or duplex, anticoagulation may be stopped or continued after 6 months in case of remaining aneurysma or irregularities of the vessel wall. Vascular surgery is not the treatment of choice for ICAD. It is only applicable in cases with recurrent symptoms of cerebral ischemia

Figure 4 Computed tomography showing left internal carotid artery (ICA) aneurysma (arrow) with partial thrombosis of its lumen (asterisk).

despite anticoagulant therapy. Prognosis is generally favorable. Studies have reported a complete or almost complete recovery in 23 to 85% of cases.¹²⁻¹⁵

Despite the high sensitivity of conventional MRI, in our case the vascular alteration was mistaken for an inflammatory reaction around the vessel. The dissection of the artery was overlooked and angiography was only performed after a surgical exploration of the field. In retrospect, surgery was not only unnecessary, but also seriously endangered our patient.

CONCLUSION

ICAD should always be included in the differential diagnosis of lower cranial nerve palsy, despite its rare causal connection. Clinical symptoms can be atypical and misleading as seen in our patient. Nevertheless, a thorough assessment including ultrasound, MRI/CT, and angiography should lead to the correct diagnosis without delay and additional risk to the patient.

REFERENCES

1. Schievink WI, Mokri B, Whisnant JP. Internal carotid artery dissection in a community. Rochester, Minnesota, 1987–1992. *Stroke* 1993;24:1678–1680
2. Giroud M, Fayolle H, André N, et al. Incidence of internal carotid artery dissection in the community of Dijon. *J Neurol Neurosurg Psychiatry* 1994;57:1443
3. Schievink WI, Mokri B, O'Fallon WM. Recurrent spontaneous cervical-artery dissection. *N Engl J Med* 1994;330:393–397
4. Bogousslavsky J, Regli F. Ischemic stroke in adults younger than 30 years of age. Cause and prognosis. *Arch Neurol* 1987; 44:479–482 Erratum in: *Arch Neurol* 1987 Aug; 44:817
5. Mokri B, Silbert PL, Schievink WI, Piepgras DG. Cranial nerve palsy in spontaneous dissection of the extracranial internal carotid artery. *Neurology* 1996;46:356–359
6. Boström K, Liliequist B. Primary dissecting aneurysm of the extracranial part of the internal carotid and vertebral arteries. A report of three cases. *Neurology* 1967;17:179–186
7. Peters M, Bohl J, Thömke F, et al. Dissection of the internal carotid artery after chiropractic manipulation of the neck. *Neurology* 1995;45:2284–2286
8. Schievink WI. Spontaneous dissection of the carotid and vertebral arteries. *N Engl J Med* 2001;344:898–906
9. Goldberg HI, Grossman RI, Gomori JM, Asbury AK, Bilaniuk LT, Zimmerman RA. Cervical internal carotid artery dissecting hemorrhage: diagnosis using MR. *Radiology* 1986;158:157–161
10. Lévy C, Laissy JP, Raveau V, et al. Carotid and vertebral artery dissections: three-dimensional time-of-flight MR angiography and MR imaging versus conventional angiography. *Radiology* 1994;190:97–103
11. Sturzenegger M, Mattle HP, Rivoir A, Baumgartner RW. Ultrasound findings in carotid artery dissection: analysis of 43 patients. *Neurology* 1995;45:691–698
12. Mokri B, Sundt TM Jr, Houser OW, Piepgras DG. Spontaneous dissection of the cervical internal carotid artery. *Ann Neurol* 1986;19:126–138
13. Ast G, Woimant F, Georges B, Laurian C, Haguenau M. Spontaneous dissection of the internal carotid artery in 68 patients. *Eur J Med* 1993;2:466–472
14. Pozzati E, Giuliani G, Acciarri N, Nuzzo G. Long-term follow-up of occlusive cervical carotid dissection. *Stroke* 1990;21:528–531
15. Sturzenegger M, Mattle HP, Rivoir A, Baumgartner RW. Ultrasound findings in carotid artery dissection: analysis of 43 patients. *Neurology* 1995;45:691–698