Acta neurol. belg., 2007, **107**, 91-93

Case reports

A carotid artery dissection presenting with dysphagia due to a dilation of upper oesophagus

M. VAES[1], E. SELLITTI[2], F. SUKKARIEH[3] and M. VANHAEVERBEEK[1]

[1]Department of Internal Medicine, [2]Department of Neurology, [3]Department of Radiology, CHU Vésale, Montigny-Le-Tilleul, Belgium

Abstract

Carotid and vertebral artery dissection is one of the major causes of ischemic stroke in young patients and diagnosing it remains a challenge for the clinician due to its variable clinical presentation. We report the case of a 57-years old man admitted to the emergency department complaining of dysphagia and a hoarse voice. Physical examination revealed a left deviation of his tong and some weakness of the left side of his soft palate. A gastroscopy and a cervical tomodensitometry revealed the presence of an important dilation of his upper oesophagus. A cerebral magnetic resonance was performed, showing en excentric high signal intensity on the left carotid artery, and the MR angiography confirmed the presence of an acute dissection on the left internal carotid artery. Our patient suffered of a left carotid artery dissection presenting with ipsilateral Xth and XIIth nerve palsies. His main symptom was a dysphagia due to a dilation of his upper oesophagus ; the pneumogastric nerve being responsible for the parasympathic innervation of the upper muscles of the oesophagus.

Key words : Carotid, dissection ; oesophagus ; dysphagia ; cranial, nerve ; palsy.

Introduction

Carotid and vertebral artery dissection is one of the major causes of ischemic stroke in young patients. While they account for about 2% of all ischemic strokes, they are associated in about 20 % of strokes in young adult patients (Ducrocq X. *et al.*, 1999), with a peak of incidence in the fifth decade (Schievink W. I., 2001).

The major challenge to the treating physician is the highly variable clinical presentation of carotid artery dissection (CAD), making the diagnosis difficult. Fortunately, improvement in investigational methods, such as magnetic resonance imaging, often helps the clinician to establish the diagnosis.

We report a case of a 57-years old man presenting with dysphagia due to a dilation of his upper oesophagus.

Case report

A 57-years old man was admitted to the emergency department complaining of a progressive dysphagia which had been evolving for fifteen days. He also noted a hoarse voice. He had no recent history of neck trauma. He was an alcoholic and smoker and his major antecedent was a mania-co-depression for which he had a treatment with lithium.

On physical examination a left deviation of his tong and some weakness of the left side of his soft palate were noted.

A gastroscopy was performed, revealing the presence of an important dilation of his upper oesophagus. The otolaryngologist discovered a left vocal cord palsy. The cervical tomodensitometry confirmed the presence of the oesophagus dilation but neither a cervical mass nor an adenopathy was observed (Fig. 1).

A cerebral tomodensitometry showed non significative ischaemic lesions.

Finally a cerebral magnetic resonance was performed. Axial T1- and T2-weighted images showed an excentric high signal intensity on the left carotid artery. On the MR angiography, an axial view showed a high-grade stenosis of 12 mm long on the left internal carotid artery, on her extracranial portion, upstream the carotid siphon. The carotid artery presented a flame-like occlusion, typical of an acute dissection (Fig. 2).

An anticoagulation therapy was started with intravenous heparin followed by oral anticoagulation with coumarines.

A Doppler ultrasound of the carotid arteries showed no evidence of atherosclerotic plaque, vascular obstruction or carotid dissection. The echocardiogram showed a postero-septal hypokinesia with a normal cardiac function.

Discussion

Dissection of the carotid and vertebral arteries usually arise from a tear in the wall of an artery,

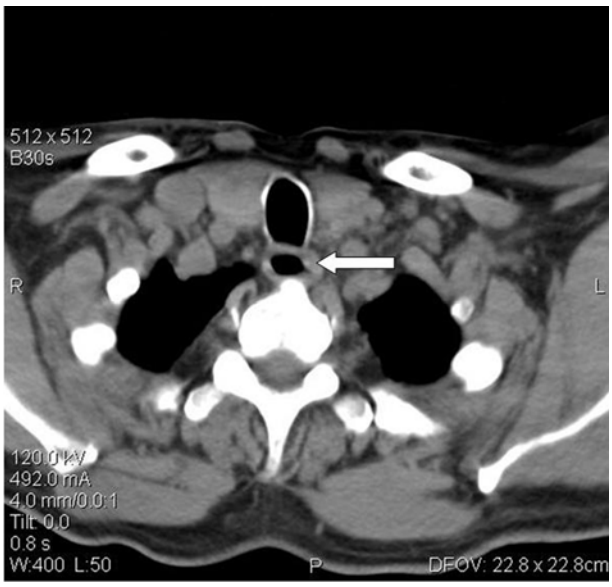

FIG. 1. — Oesophagus dilation on cervical tomodensitometry (see white arrow).

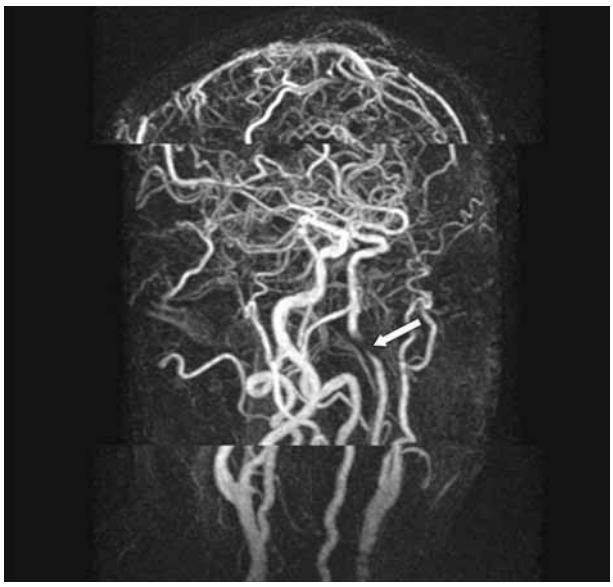

FIG. 2. — MR angiography : a flame-like occlusion of the internal carotid artery upstream the carotid siphon (see white arrow).

leading to a collection of blood between the layers of the artery. This leads to the formation of an intramural hematoma, the so-called false lumen. Whether the primary site of dissection is the intima (with arterial blood penetrating the wall) or the media (leading to a primary intramural hematoma reconnecting later with the true lumen) is not well established (Schievink W. I., 2001 ; Thanvi B. *et al.* ; 2005). However, the result is either stenosis or aneurismal dilatation of the vessel.

Several factors have been identified as triggers or causes of CAD. An underlying arteriopathy, as observed in Ehlers-Danlos syndrome (Schievink W. I., Michels V. V. *et al.*, 1994) or Marfan's syndrome (Schievink W. I., Björnsson J. *et al.*, 1994) can be associated to CAD, although in practice they are rarely found. CAD has also been reported in association with fibromuscular dysplasia or cystic medial necrosis (Schievink W. I. *et al.*, 1994).

Precipitating events leading to CAD include all the movements of hyper-extension or rotation of the neck (yoga, …), neck or head trauma in motor vehicle accidents, chiropractic manipulation (Leys D. *et al.*, 1995 ; Norris J. W. *et al.*, 2000 ; Dziewas R. *et al.*, 2003).

Atherosclerosis appears to be distinctly uncommon in patients with dissection, although the relation between cardio-vascular risk factors like tobacco use or hypertension and CAD has not been systematically evaluated (Mokri B. *et al.*, 1986).

The major difficulty of diagnosing CAD is due to the high variety of clinical presentations. Some dissections remain asymptomatic, some present isolated subtle manifestations, and the classic triad including headache or neck pain, a partial Horner's syndrome and cerebral or retinal ischemia, is rarely present.

The neck pain often develops on one side of the neck and usually there is an ipsilateral headache (Silbert P. L. *et al.*, 1995). Our patient complained of headache during his hospitalisation. The pain started in the neck and involved the entire left hemicranium. Neck pain or headache is usually the initial manifestation of CAD and the median time to the appearance of other symptoms is four days (Silbert P. L. *et al.*, 1995). Our patient didn't mention any headache on his arrival at the emergency service complaining that he had suffered of dysphagia for fifteen days, so it was not a typical presentation.

Although oculosympathetic palsy (Horner's syndrome) has long been recognized as a typical manifestation of CAD, it is found in less than half of patients (Mokri B. *et al.*, 1979 ; Hart R. G. *et al.*, 1983 ; Silbert P. L. *et al.*, 1995).

Cranial nerve palsies can be detected in about 12% of patients with CAD (Mokri B. *et al.*, 1996).

Various combinations of nerve palsies have been described, although the lower cranial nerves, particularly the hypoglossal nerve, are the most commonly affected.

Two different mechanisms have been described to explain cranial nerve palsies. The first one is a compression of the expanded artery. Indeed, the cervical parapharyngeal space contains the four lower cranial nerves, situated between the internal carotid artery medially and the jugular vein distally. Those nerves may be involved in the expanded wall haematoma that can nearly triple the carotid diameter. The XIIth nerve is the closest to the internal carotid, which could explain why it is the most frequently involved (Guy N. *et al.*, 2001). The second mechanism is an impairment of the nutrient arterial supply to the related nerve, resulting of distal embolisation, pressure gradient changes in

collateral supply, or impairment of the nutrient vessel by the dissection (Guidetti D. *et al.*, 2001).

Our patient presented a palsy of the nerves X and XII. The particularity of this case is the Xth nerve palsy, resulting in a dilatation of the upper oesophagus. Indeed, the pneumogastric nerve is responsible for the parasympathic innervation of the upper muscles of the oesophagus.

So the major difficulty for the physician is the wide variety of clinical presentations associated to CAD, the lack of specific symptoms and the low prevalence of the disease.

The clinician establishes various hypotheses of diagnosis according to the patient's clinical manifestations, his risk factors and the frequency of diseases. These hypotheses are also influenced by the physician's knowledge and personal experience. In this case, the ethylo-tabagic patient presenting a dysphagia and a palsy of the hypoglossal nerve oriented first our diagnosis towards a cervical neoplasia with cerebral metastasis. Fortunately angiographic RMN revealed the artery dissection.

Conventional angiography has long been the gold-standard for diagnosing CAD but magnetic resonance techniques are replacing it (Kasner S. E. *et al.*, 1997).

Doppler ultrasonographic techniques can be usefull to detect an abnormal flow but the site of dissection is often not seen (Sturzenegger M. *et al.*, 1995).

Anticoagulation with intravenous heparin followed by oral warfarin has been recommended to prevent thrombo-embolic complications, though the benefit of an anticoagulation has not been evaluated with randomized trials (Lyrer P. *et al.*, 2003).

Conclusion

We reported a case of CAD presenting dysphagia as main symptom of a palsy of the Xth cranial nerve resulting in a dilation of the upper oesophagus.

Diagnosing CAD remains a challenge for the clinicians, due to its variable presentations. Headache or neck pain, and cranial nerve palsies, especially the last ones, can lead to think about it. It is crucial to recognize atypical findings and to perform an accurate and prompt diagnostic evaluation. Angiographic RMN remains the best non invasive procedure to reveal the dissection.

REFERENCES

Ducrocq X., Lacour J. C., Debouverie M., Bracard S., Girard F. *et al.* Accidents vasculaires cérébraux ischémiques du sujet jeune : étude prospective de 296 patients âgés de 16 à 45 ans. *Rev. Neurol. (Paris),* 1999, **155** : 575-582.

Dziewas R., Konrad C., Dräger B., Evers S., Besselmann M. *et al.* Cervical artery dissection—clinical features, risk factors, therapy and outcome in 126 patients. *J. Neurol.*, 2003, **250** (10) : 1179-84.

Guidetti D., Pisanello A., Giovanardi F., Morandi C., Zuccoli G. *et al.* Spontaneous carotid dissection presenting lower cranial nerve palsies. *J. Neurol. Sci.*, 2001, **184** (2) : 203-7.

Guy N., Deffond D., Gabrillargues J., Carriere N., Dordain G. *et al.* Spontaneous internal carotid artery dissection with lower cranial nerve palsy. *Can. J. Neurol. Sci.*, 2001, **28** (3) : 265-9.

Hart R. G., Easton J. D. Dissections of cervical and cerebral arteries. *Neurol. Clin.*, 1983, **1** : 155-182.

Kasner S. E., Hankins L. L., Bratina P., Morgenstern L. B. Magnetic resonance angiography demonstrates vascular healing of carotid and vertebral artery dissections. *Stroke*, 1997, **28** : 1993-1997

Leys D., Moulin Th., Stojkovic T., Begey S., Chavot D., DONALD Investigators. Follow-up of patients with history of cervical artery dissection. *Cerebrovasc. Dis.*, 1995, **5** : 43-49.

Lyrer P., Engelter S. Antithrombotic drugs for carotid artery dissection. *Cochrane Database Syst. Rev.*, 2003, (3) : CD000255.

Mokri B., Silbert P. L., Schievink W. I., Piepgras D. G. Cranial nerve palsy in spontaneous dissection of the extracranial internal carotid artery. *Neurology*, 1996, **46** : 356-359.

Mokri B., Sundt T. M. Jr., Houser O. W. Spontaneous internal carotid dissection, hemicrania, and Horner's syndrome. *Arch. Neurol.*, 1979, **36** : 677-680 .

Mokri B., Sundt T. M. Jr., Houser O. W., Piepgras D. G. Spontaneous dissection of the cervical internal carotid artery. *Ann. Neurol.*, 1986, **19** : 126-138.

Norris J. W., Beletsky V., Nadareishvili Z. G., Canadian Stroke Consortium. Sudden neck movement and cervical artery dissection. *CMAJ*, 2000, **163** : 38-40.

Schievink W. I. Spontaneous dissection of the carotid and vertebral arteries. *N. Engl. J. Med.*, 2001, **344** (12) : 898-906.

Schievink W. I., Björnsson J., Piepgras D. G. Coexistence of fibromuscular dysplasia and cystic medial necrosis in a patient with Marfan's syndrome and bilateral carotid artery dissections. *Stroke*, 1994, **25** : 2492-2496.

Schievink W. I., Michels V. V., Piepgras D. G. Neurovascular manifestations of heritable connective tissue disorders : a review. *Stroke*, 1994, **25** : 889-903.

Silbert P. L., Mokri B., Schievink W. I. Headache and neck pain in spontaneous internal carotid and vertebral artery dissections. *Neurology*, 1995, **45** : 1517-1522.

Sturzenegger M., Mattle H. P., Rivoir A., Baumgartner R. W. Ultrasound findings in carotid artery dissection : analysis of 43 patients. *Neurology*, 1995, **45** : 691-698.

Thanvi B., Munshi S. K., Dawson S. L., Robinson T. G. Carotid and vertebral artery dissection syndromes. *Postgrad. Med. J.*, 2005, **81** (956) : 383-8.

Mélanie Vaes, M.D.,
Rue Colonel Chaltin 37/1,
B-1180 Uccle (Belgium).