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Unilateral oculomotor nerve palsy caused by a basilar artery dolichoectasia

Paralysie unilatérale du nerf oculomoteur commun causée par une dolichoectasie du tronc basilaire

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Résumé

L'atteinte isolée de l'un des nerfs crâniens oculomoteurs due à une dolicho-ectasie vertébro-basilaire est rare. Nous rapportons le cas d'une patiente ayant présenté une diplopie horizontale d'installation aigue liée à une paralysie partielle du nerf oculomoteur commun gauche. La neuro-imagerie a mis en évidence une anomalie vasculaire de l'artère basilaire suggérant une dolicho-ectasie à proximité de la partie interne du pédoncule cérébral gauche. La patiente a complètement et spontanément récupéré après un mois. Ce cas illustre une étiologie rare de paralysie partielle du nerf oculomoteur commun épargnant la pupille.

Mots-clés : Paralysie du nerf III, nerf oculomoteur, dolicho-ectasie vertébro-basilaire

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Summary

Isolated dysfunction of the third cranial nerve due to vertebrobasilar dolichoectasia is uncommon. We report a case of a patient who presented with recent onset of horizontal diplopia due to left partial third nerve palsy. Neuroimaging revealed a vascular abnormality in the basilar artery suggesting a dolichoectasia next with a leftward deviation to the internal side of the left cerebral peduncle. The patient fully and spontaneously recovered after one month. This case illustrates a rare etiology of a partial oculomotor nerve palsy sparing the pupil.

Keywords: Third nerve palsy, Oculomotor nerve, Vertebro-basilar dolichoectasia

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Abbreviations
VBD: Vertebrobasilar dolichoectasia
MRI: Magnetic resonance imaging
CA: conventional angiography

Introduction

Vertebrobasilar dolichoectasia (VBD) is a rare condition characterized by an abnormal dilation, elongation, widening and tortuosity of the vertebrobasilar arteries (1). It is usually an asymptomatic condition, but when symptomatic, can reveal with 2 types of symptoms: vascular events (including ischemic or hemorrhagic) or compression of neighboring structures (2-3). Cranial nerves dysfunction is usually caused by compression by the enlarged vessels and is usually revealed by progressive symptoms (2). In this article, we describe a patient who developed an isolated left third cranial nerve palsy caused by its compression by a dolichoectatic basilar artery as it emerges from the internal side of the left cerebral peduncle.

A 50-year-old woman presented to the emergency room complaining of a recent onset horizontal diplopia, with unusual left unilateral pulsating headache of mild intensity, without any other symptoms. Past medical history was marked by chronic tension-type headache without history of diabetes neither hypertension. Neurological examination found a rightgaze and downgaze palsy of the left eye, without abnormal pupillary response, corresponding to left partial third nerve palsy (figure 1).

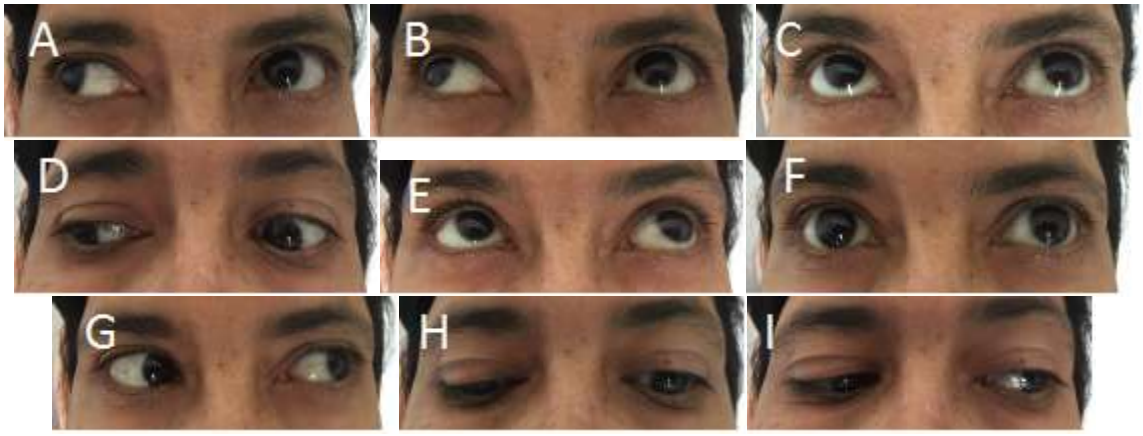

Figure 1. Cardinal positions of gaze showing a rightgaze (A) and downgaze (H) palsy of the left eye

She had no other neurological deficit. Ophthalmologic examination was unremarkable. Magnetic resonance imaging (MRI) revealed the existence of a leftward deviation of the basilar artery with a very close relationship to the internal side of the left cerebral peduncle and without any parenchymal abnormality (figures 2 and 3).

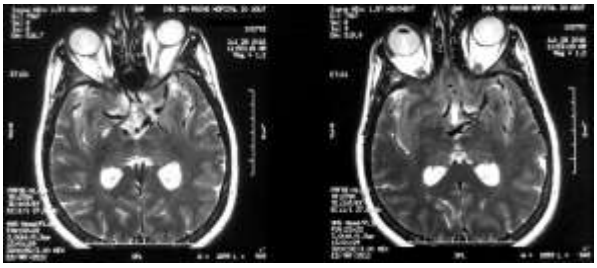

Figure 2. T2-weighted images showing a leftward deviation of the basilar artery (basilar artery dolichoectasia) (blue arrow) and mild hydrocephalus (red arrow)

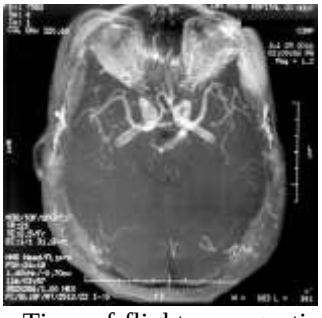

Figure 3. Time-of-flight magnetic resonance angiogram showing an elongated basilar artery (basilar artery dolichoectasia) (blue arrow)

Gradient echosequence did not show any evidence of hemorrhage. Conventionnal angiography (CA) confirmed the diagnosis of basilar artery dolichoectasia (figure 4).

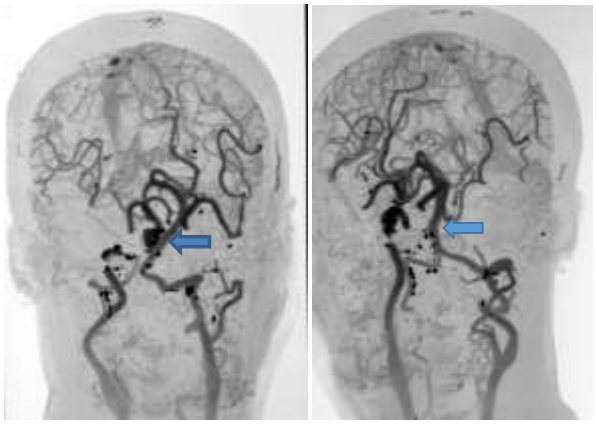

Figure 4. Conventional angiography images showing an elongated basilar artery (basilar artery dolichoectasia) (blue arrow)

Glucose level, serum lipid levels and inflammation markers (sedimentation rate and CRP) were within normal limits. The patient recovered fully and spontaneously after one month. She was recommended to monitor her blood pressure to detect hypertension as it can be a deleterious comorbidity for the vessels.

Discussion

Epidemiology

The incidence of intracranial dolichoectasia varies between 0.06–5.8 percent (4). Dolichoectasia most commonly involves the vertebral and basilar arteries (4-5). It is a clinical disorder based on established radiological (Computed Tomography or MRI) criteria including a basilar artery >4.5 mm in diameter, deviation of any portion of both arteries more than 10 mm from its shortest expected course, a basilar artery length >29.5 mm or intracranial vertebral length >23.5 mm (4-5).

Pathogenesis/Risk factors

The primary pathophysiologic mechanism of dolichoectasia is an aberrant vascular remodeling and abnormal connective tissue within the arterial wall due to an imbalance between matrix metalloproteinases and antiprotease activity in the connective tissue (1-3). The association of VBD with enlargement of the descending thoracic aorta and coronary arteries suggest that it may be part of a systemic vasculopathy (2). The main risk factors of this entity are: older age, male sex, previous stroke, diabetes, hypertension, smoking and alcohol abuse. Moreover, low renal function and age seem to be specifically associated with greater dilatation in the posterior circulation. Other factors include autosomal dominant polycystic kidney disease, congenital or acquired collagen disease (Ehlers-Danlos and Marfan syndromes), inherited metabolic disorders (Fabry disease) and hereditary brain malformations (1,3). The only factor found in our case was advanced age.

Diagnosis

In some patients, VBD is an incidental radiological anomaly. But sometimes, it produces neurological signs and symptoms related to brainstem ischemia, impaired cerebrospinal fluid dynamics or a combination of these mechanisms. The progressive compression of the cranial nerve roots has also been reported and most commonly involves the trigeminal and facial nerves (3-6). It may also have rare clinical presentations, such as painful tic, neuralgia, tinnitus, vertigo, headache, migraine and central sleep apnea (4). Although the diagnosis was confirmed in our case by CA, non-invasive neuroimaging studies such as magnetic resonance angiography and particularly Computed Tomography Angiography are sufficient to establish the diagnosis of VBD (6).

Therapeutic options

There is currently no effective treatment for VBD itself because of the location and the important major arterial and perforating branches that supply critical structures. Some surgical techniques with inconsistent outcomes and morbidities such as clip reconstruction, microvascular decompression with vessel repositioning, surgical reduction of basilar artery diameter have been attempted. However, the benefits of surgery remain to be established. Some reports indicate that poor outcome and mortality rates surpasses 16% (1), with brainstem infarction as the most common complication (up to 22% of cases). Endovascular interventional treatments, such as flow diversion using stents with or without coils are better options because of their safety. Overall, treatment is usually reserved for symptomatic cases of VBD. The treatment of VBD-related stroke is controversial because conventional doses of antiplatelet and anticoagulant drug therapy may increase the risk for intracranial bleeding; hence these drugs should be prescribed with caution (2). For compressive nerves symptoms such as in our patient, the most effective treatment is reported to be microvascular decompression by interposing synthetic implants between the culprit vessel and the nerves (2). Sometimes, the symptoms can regress spontaneously similar to the clinical course observed in our patient (6).

Prognosis

The long-term prognosis of patients with VBD depends on the severity of the dolichoectasia at diagnosis, the diameter of the basilar artery, the height of bifurcation, and its evolutionary characteristics (2). Large basilar artery diameter, stroke at presentation and posterior circulation

dolichoectasia are all associated with early and higher than average stroke recurrence rates (1). The cumulative incidence of dolichoectasia related stroke after 1 year, 5 years and 10 years follow up is 2.7, 11.3 and 15.9%, respectively (1). The risk of stroke from thrombotic occlusion of the basilar artery or its branches or embolization to the distal vascular territories is overall 52–77%, thus VBD is associated with a significant morbidity and mortality (5).

Isolated dysfunction of one of the ocular motor cranial nerves is uncommon in the case of VBD; impairment of the abducens and IVth nerve has been described (5-6). Involvement of the third cranial nerve has also been reported in a few clinical cases (7-11).

Conclusion

The reported case illustrates a rare etiology of a partial oculomotor nerve palsy sparing the pupil. Therefore, clinicians should consider VBD as a possible cause of an isolated third nerve palsy. Patients should be aware that even if the nerve palsy resolves spontaneously as in our patient, the long-term prognosis is not always benign as other cranial nerve palsies may emerge as well as significant neurological dysfunctions due to ischemic stroke (5).

Declaration of Conflicting Interests

The Authors declare that there is no conflict of interest

Author's contribution

Data collection and interpretation: Hicham El Otmani; Drafting the article: Vicky Fotso; Critical revision of the article: Salma Bellakhdar, Mohamed Abdoh Rafai and Bouchra El Moutawakil; all authors approved the final version.

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