

ISOLATED PATHETICK NERVE PARESIS BY COMPRESSION FROM A DOLICHOECTATIC BASILAR ARTERY

Case report

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ABSTRACT - The article describes a woman with 53 year-old that has presented diplopia when looking down and adopted a tilted head position in order to read for the last six months. The neuro-ophthalmic examination shows isolated right superior oblique muscle paresis. The magnetic resonance and the angioresonance show the dolichoectatic basilar artery compressing the right ventral lateral brainstem.

KEY WORDS: trochlear nerve, dolichoectasy, basilar artery, ophthalmoplegia, compression.

Paresia isolada do nervo patético por compressão da artéria basilar dolicoectásica: relato de caso

RESUMO - O artigo descreve uma mulher de 53 anos que há seis meses apresenta diplopia ao olhar para baixo, além de adotar postura inclinada da cabeça para ler. O exame neuro-oftalmológico revelou paresia isolada do músculo oblíquo superior direito. Os exames de ressonância magnética e angioresonância evidenciaram dolicoectasia da artéria basilar, que comprimia a face ventral lateral direita do tronco cerebral.

PALAVRAS-CHAVE: nervo troclear, dolicoectasia, artéria basilar, oftalmoplegia, compressão.

The term dolichoectasia is derived from "dolichos", meaning elongation and "ectasia", meaning dilatation¹. Dolichoectasia of the intracerebral vessels is a rare disorder that affects the large arteries at the base of the brain^{2,4}, most commonly the basilar artery^{3,5}. In 1.7% of the general population, that is the only affected artery⁶. In a retrospective study reviewing 1440 magnetic resonance angiographies with the radiological diagnosis of vertebrobasilar dolichoectasia, Obogu et al.³, suggested specific parameters for defining this condition. Those were: basilar artery (BA) or vertebral artery (VA) diameter > 4.5 mm or deviation of any portion >10 mm from the shortest expected course, and BA length >29.5 mm or intracranial VA length >23.5 mm. Compression by a dolichoectatic basilar artery (DBA) has already been involved in ocular motor nerve paresis⁶. Although it is less common than the compromising of the oculomotor or abducens nerves, the trochlear

nerve may be involved by mechanical compression caused by dolichoectasia⁷.

The purpose of this case report is to present an unusual association of isolated paresis of the trochlear nerve with a DBA.

CASE

Six months ago, a 53 year-old hypertensive scholar group teacher started feeling persistent double vision when looking down. She started reading and writing with her head bent to the left. Even though she made orthoptic exercises for two months, her condition did not improve. She denies headache, vomiting, vertigo, cranial trauma, and any motor or sensitive symptom. She is not diabetic and does not use tobacco or consume alcohol.

On clinical examination she was lucid, oriented, and helped with the examination. Blood pressure (checked in both arms) was 240x120 mmhg while seated. Neuro-ophthalmic examination shows that she had no ptosis, eyelid twitch, fluttering eye movements, or eyelid fatigue after

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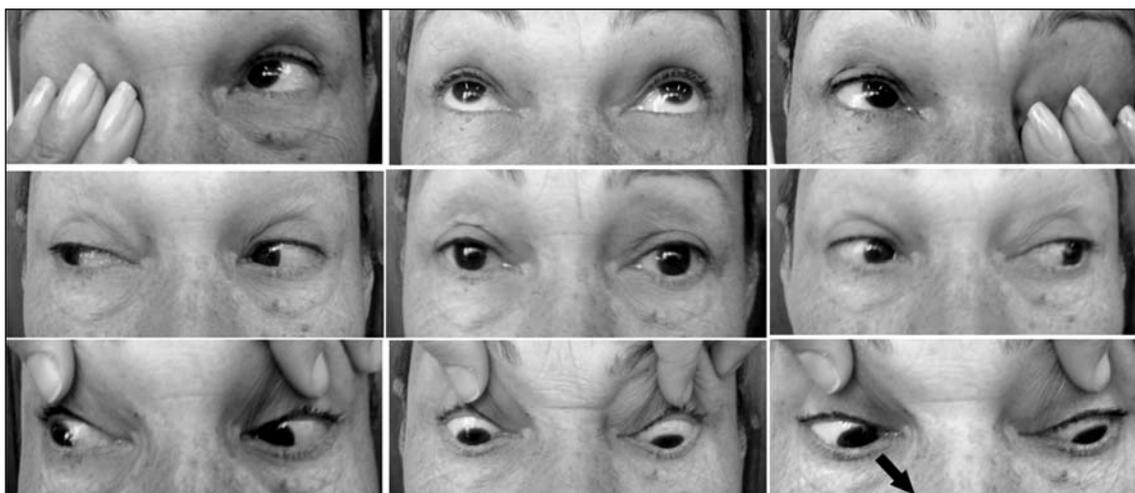


Fig 1. Extraocular movements in 9 cardinal positions. Limitation of the right superior oblique muscle on gaze down and left (arrow).

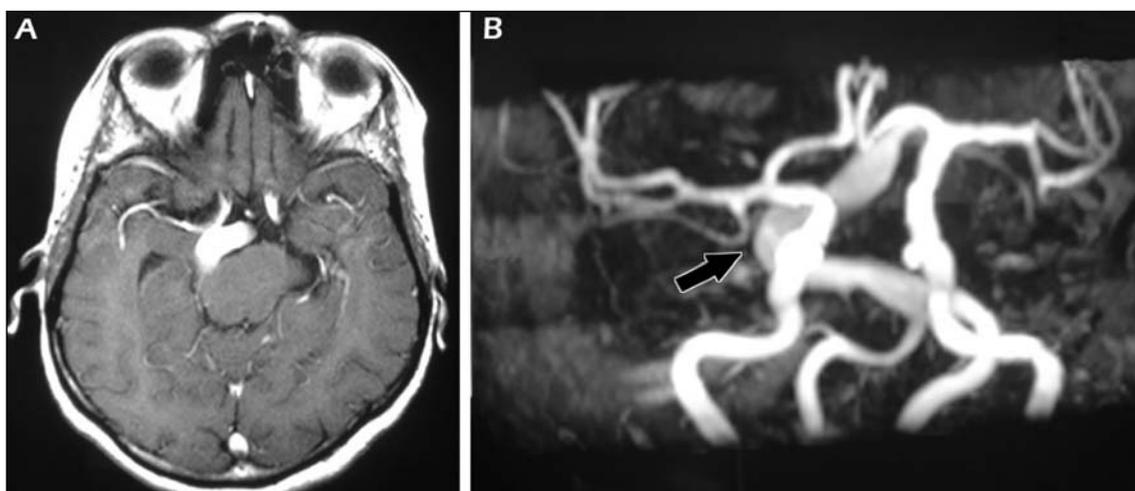


Fig 2. (A) T1W axial magnetic resonance imaging following gadolinium infusion shows a markedly enlarged basilar artery with upper pons compression. (B) Magnetic resonance arteriography shows a dolichoectatic basilar artery moved more than 10 mm to the right (arrow).

sustained upgaze. Visual acuity, color vision, pupil size, and reactivity were normal. Except for a few Gunn's crossing phenomena, the *fundus oculi* were normal. She complained about vertical diplopia when asked to read or write, and bent her head to the left (ear-shoulder position) in order to reduce visual discomfort. The extra ocular motility examination showed paresis of the right superior oblique muscle (Fig 1), confirmed by the Bielschowsky head-tilt test. Magnetic resonance image (MRI) and magnetic resonance angiography (MRA) showed DBA with brainstem compression (Fig 2A and B). The neurosurgery team discarded the possibility that microvascular decompression would eliminate the mild symptoms. Furthermore, the patient refused the risks of major surgery. She maintains her blood pressure under control with the aid of internal medical visits.

The patient has formally given authorization for the publication of this case report

DISCUSSION

By 1664, Thomas Willis accurately described the fourth nerve: "*We call these nerves the Pathetick nerves of the eyes . . . the proper office of these is to move the eyes pathetically, according to the force of the passions and instinct of nature*"⁸.

Isolated trochlear nerve palsy has been described as a result of the most diverse causes, such as: diabetes ischemic neuropathy⁹, traumas¹⁰, infectious diseases, including meningitis⁸, tumors¹¹, demyelinating disease¹², aneurysm¹³, strokes¹⁴, and complication of intracranial surgery¹⁵.

DBA may be asymptomatic, but in 6% of this patients it may cause clinical manifestations with

acute onset (stroke or subarachnoid hemorrhage) or with chronic progressive presentation (cranial nerve palsy, compression of the midbrain, or obstructive hydrocephalus)^{6,16}.

Although rare, a possible site for vascular involvement of the fourth nerve would be mechanical compression as it courses towards the ventrolateral aspect of the pons, in the free border of *tentorium cerebelli*, by a dolichoectatic dilatation of a sclerotic vessel or a congenital aneurysm⁷. This was not found in larger series of pathetic paralysis, like in Rucker's¹⁷ 67 cases, and in only one of Burger's 33 cases⁷ when an aneurysm (not dolichoectatic artery) was found compressing the trochlear nerve.

The pathophysiology of DBA is unknown. On autopsy, the pathologic findings are rarefaction of the elastic tissue in the media with degeneration of the internal elastic lamina. Sometimes, there is an intraluminal thrombus or an atherosclerotic plaque in the elastic artery⁵. The dolichoectatic arteries have an abnormally large external diameter with a thin arterial wall¹⁶.

Some years ago, Keane⁸ published a study concerning 215 patients with uni or bilateral palsy of the trochlear nerve and made a review on this issue. Traumas were a cause in more than half of the patients, producing 90 (51%) of the unilateral and 23 (59%) of the bilateral fourth nerve palsies. Although there were many described causes, there is no mention of DBA being the causing agent.

Because of the unique visual symptom, vertical diplopia, which is worse on gaze down or near than up or distant, most of the patients look initially for an ophthalmologist's office⁸. The clinical diagnosis of superior oblique weakness is based on the presence of three main aspects: first, vertical diplopia is maximal when the affected (higher) eye is turned down such as when descending a flight of stairs⁹ or reading lying down or seated at a table, like this teacher.

Second, a characteristic compensatory head posture for writing or reading that our patient adopted. Like in other patients with the same problem, ours compensated for the torsional deviations of the eyes by tilting her head to the left, the horizontal deviations by turning her face to the right, and for vertical deviations by lowering her chin in order to facilitate fusion and to minimize double vision⁷.

The third aspect is the Bielschowsky head-tilt test⁹. We asked our patient to demonstrate, using her finger, the distance between two images when her head was tilted to right, and then to the left. There was greater separation of images when she tilted her head to the right.

In conclusion, isolated paresis of the trochlear nerve is a rare condition, and even rarer when resulting from DBA compression. The neuro-ophthalmic examination is characteristic, and the combination of MRI and MRA is an effective and noninvasive means of identifying a vascular loop as the cause of the ophthalmic paresis.

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