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# **Dolicho-Vertebrobasilar Abnormality and Migraine-Like Attacks**

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## **Key Words**

Dolicho-vertebrobasilar anomalies • Trigeminal nerve • Migraine • Magnetic resonance imaging • Magnetic resonance angiography

## Abstract

This is a study of two children with neuroradiologically diagnosed dolichobasilar anomaly: in both cases, the diagnosis was based on magnetic resonance imaging and magnetic resonance angiography findings. We examined the first child (R., aged 6 years and 8 months) following four episodes of intense pain in the left orbit, lasting between 10 and 30 min and accompanied by photophobia and vomiting. The second child (S., aged 9 years and 2 months), suffering from Marfan's syndrome, was examined following recurrent episodes of intense pain that started above the left orbit, moving to the homolateral orbit after a few minutes. These episodes, which lasted between 20 and 30 min, took place mainly at night and were accompanied by phono-photophobia and vomiting. In both cases, the pain attacks can be interpreted as a result of compression of the root of the trigeminal nerve caused by an arterial malformation.

## Introduction

Anomalies in the diameter and course of the basilar artery are not infrequent in adults and can be found isolated or as part of more complex malformations of the cerebral arteries [1]. Many synonyms have been used to describe these conditions: fusiform aneurysm, S-shaped aneurysm, tortuous or elongated basilar artery, basilar artery dilatation, vertebrobasilar dolichoectasia and megadolichobasilar artery (MDBA). Smoker et al. [2] have shown that the elongation or twisting of the vertebrobasilar system without associated ectasia can give rise to neurological signs and symptoms. They therefore propose a differentiation between dolicho-vertebrobasilar and vertebrobasilar dolichoectasia, and underline that patients with dolicho-vertebrobasilar artery often present with isolated involvement of a single cranial nerve, whereas patients with vertebrobasilar dolichoectasia present with multiple neurological deficits. From the pathogenic point of view, these conditions were considered secondary either to arteriosclerotic alterations in the vessel, or, more convincingly, to structural anomalies in the vessel wall that are probably congenital and linked to an unknown alteration in collagen metabolism [3, 4]. To support this hypothesis, we cite the association of vascular cerebral and extracerebral malformations with Marfan's syndrome, Ehlers-Danlos syndrome, pseudoxantoma elasticum [5] and with other less well-defined connective tissue disorders [6]. In the majority of cases, symptoms

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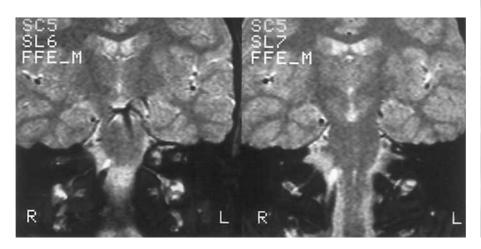
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**Fig. 1.** Coronal  $T_2$ -weighted images (TR 500, TE 19, FA 15). The loop of the basilar artery. A small vessel, very close to the left fifth cranial nerve, probably the AICA.



**Fig. 2.** Three-dimensional TOF-MRA: the basilar artery forms a loop from right to left; the right vertebral artery is also elongated, while the left one is missing.

begin between the ages of 50 and 60, occurring only extremely rarely in children. In three pediatric cases, described in the literature, dolichoectasia of the cerebral arteries was present in the form of fusiform aneurysm [3, 7, 8]; in another case it appeared as an incidental feature of a complex vascular cerebral anomaly [4].

Clinically, anomalies are discovered mainly as a result of the compression by the dolichoectatic artery of neighboring structures, or following ischemic vascular accidents. In terms of MDBA, Masson and Cambier [9] subdivide the symptoms into three groups: ischemic symptoms, symptoms of compression of the cranial nerves and pseudotumoral symptoms. In rare cases, intracranial hemorrhage can also be found. Nevertheless, the polymorphous nature of the clinical manifestations [1, 2, 7, 10–14] makes it difficult to establish precise diagnostic criteria. Neuroradiology does, however, appear to play an important role [1, 2, 11, 15].

In this article, we describe two pediatric patients with a neuroradiological picture suggesting dolichobasilar artery and a clinical history of recurrent pain attacks that have notable similarities.

#### **Case Reports**

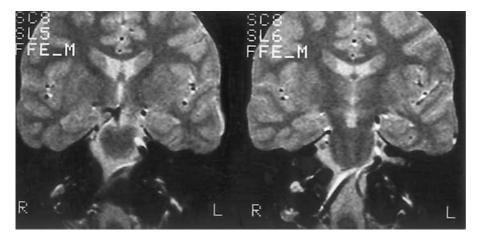
Case 1

R., male, was first examined at the age of 6 years and 8 months. His family history was negative for headache. His psychomotor development was normal, but the patient had always moved clumsily and awkwardly and had difficulty running. The patient was admitted to our department following four sudden attacks of extremely intense pain in the space of 8 months. The pain was located in and above the left orbit, accompanied by intense photophobia and vomiting; the symptoms lasted between 10 and 30 min and were followed by sleep for 2–3 h with complete remission. The patient also presented 'electric-shock'-type attacks in the same location, lasting for a few seconds and with a frequency ranging from a few to multiple daily episodes. Two further attacks were reported, lasting 10–20 s and during which R. had a sudden sensation of discomfort, loss of consciousness and fixed gaze. The attacks were followed by confusion and amnesia and were diagnosed as temporal lobe epileptic seizures.

On admission, the neurological examination revealed mild hypotonia in the arms with weak deep-tendon reflexes and mild problems in the coordination test; no trigger points were found. The following examinations were carried out: blood chemistry evaluations, EMG, ENG, brainstem auditory evoked potentials and somatosensitive evoked potential, ophthalmological examination and blink reflex, all giving normal results. EEG: Focal sharp waves in short bursts in the left parieto-occipital region; during drowsiness generalized spikes and waves were present. EEG after sleep deprivation: During drowsiness a 5-second sequence of spikes and waves was present; upon awakening, three slow bilateral paroxysmal complexes were recorded. Brain magnetic resonance imaging (MRI): The coronal T<sub>2</sub>weighted images (TR 500, TE 19, FA 15) showed a loop of the basilar artery and a small vessel, probably the AICA, very close to the left fifth cranial nerve (fig. 1). The examination was completed by magnetic resonance angiography (MRA) that confirmed the extent of the dolichobasilar abnormality and also revealed increased length and tortuosity of the right vertebral artery. However, this investigation was unable to detect the left vertebral artery which may have been missing or hypoplasic (fig. 2).

Treatment with carbamazepine (400 mg/day) was started, and a gradual reduction of symptoms was observed. From the time of the first observations (November 1991) until July 1992, no further

Vertebrobasilar Abnormality in Two Children Eur Neurol 1999;41:10-14



**Fig. 3.** Coronal T<sub>2</sub>-weighted images (TR 500, TE 19, FA 15). The loop of the basilar artery in contact with the left trigeminal nerve (the black and white appearance of the arteries is due to pulsation artefact).



**Fig. 4.** Three-dimensional TOF-MRA: the basilar artery is elongated, forming a large left-convex bend.

attacks were reported. In July 1992 and 1994, R. suffered two attacks similar to those described above. Following an increase in the dosage of carbamazepine to 500 mg/day, the symptoms disappeared.

#### Case 2

S., male, was first examined at the age of 9 years and 2 months. There was no history of headache in the family. The child had a normal psychomotor development. From the age of 3 months until 3 years old, he suffered many episodes of febrile convulsions. The child was admitted to our department following repeated sudden attacks of intense pain over a period of 6 months. These attacks were localized in the left orbit; the symptoms lasted between 20 and 30 min. The attacks usually occurred at night, waking the child, and they were accompanied by phono-photophobia and vomiting which did not, however, relieve the symptoms.

On admission, the *general medical examination* revealed marfanoid habitus characterized by abnormal length of the upper limbs compared with the torso and very long, thin fingers and toes; pectus excavatum, scapular asymmetry and scoliosis were also found. The *neurological examination* revealed a joint laxity in the upper and lower limbs. *Blood tests* were normal. The following investigations were also performed:

Ophthalmological examination: Amblyopia from uncorrected refractive fault (bilateral astigmatism); crystalline lens otherwise normal. *Echocardiogram:* Prolapsed but not insufficient mitral valve and asymptomatic aortic dilatation (30.5 mm diameter). *EEG:* Normal background activity; spikes of low amplitude sometimes followed by slow waves in occipitotemporal regions. *Brain MRI:* The coronal T<sub>2</sub>-weighted images (TR 500, TE 19, FA 15) showed a loop of the basilar artery in contact with the left trigeminal nerve (fig. 3). The examination was completed with *MRA* carried out with TOF technique with three-dimensional imaging for the study of Willis' polygonal vessels. The basilar artery appeared elongated and had a twisted shape with a large left-convex bend; the top of the artery was normal corresponding with the interpeduncu-

lar cistern. The other vessels of Willis' polygon were of normal size and shape (fig. 4).

Treatment with carbamazepine (200 mg/day) was started and led to a significant improvement, with complete remission of the pain attacks within 2 months.

#### Discussion

The medical literature contains very few examples of dolichoectasia of the cerebral arteries diagnosed in children and adolescents [3, 4, 7, 8]. From the pathogenic point of view, the reporting of cases with precociously expressed symptoms is particularly valuable because this supports the hypothesis that there exists a cause other than arteriosclerosis for dolichomegalia in the cerebral arteries [1]. Read and Esiri [8] report that an autopsy revealed a defect in the internal elastic lamina. This anatomopathological finding agrees with the defects in the internal elastic lamina and deficits of the reticular fibers in the muscular stratum found by Hegedus [16]. According to this author, an alteration in the development of the smooth muscle cells during the embryonic period could be responsible for the cases of dolichoectatic intracranial arteries in which symptoms are precociously manifested. This theory would be strengthened by the presence of congenital bone malformations or Marfan's syndrome [5]. Our two cases prompt further reflection on the interpretation and correct clinical classification of the symptoms, as well as on the possible pathogenic link between pain attacks and vascular abnormalities.

Eur Neurol 1999;41:10-14

Zambrino/Berardinelli/Martelli/Vercelli/ Termine/Lanzi

In both our cases, the features of the pain attacks have some similarities with migraine without aura: the site of the pain, the photophobia and the vomiting. But some characteristics, such as the suddenness of the attack, the fact that the pain is immediately of high intensity, the relatively brief duration and the constancy of the localization of the attacks, are more similar to episodic cluster headaches, whose association with vascular endocranial abnormality has been noted [12]. Contrary to this hypothesis, however, is the young age of the patients and the absence of symptoms of autonomic dysfunction. In adult subjects, headache is considered as a symptom associated with MDBA without its characteristics being more closely specified (except in a case described by Nishizaki et al. [17] in which the headache was associated with dizziness and vomiting and in another described by Staikow and Mattle [13] which presented the clinical picture of an exertional headache). In one case [10], a correlation is reported between ectasia of the basilar artery and unusual facial pain.

R. also presented episodes of impairment of consciousness that can be interpreted, according to the neurophysiological data, as temporal lobe epileptic seizures. In the second case, we found multifocal EEG abnormalities in the absence of symptoms. In the literature, epilepsy is presented as one of the symptoms associated with MDBA in adult patients: Yu et al. [14] report a case of temporal lobe epilepsy and Gautier et al. [1], in a survey of 204 cases, found epilepsy in 10. Nevertheless, the relationship between epilepsy, EEG anomalies and vascular malformations remains unclear, and we, too, were unable to find a convincing explanation in relation to our patients.

In both cases, the hypothesis of a possible trigeminal neuralgia must still be considered. The involvement of the trigeminal nerve is one of the commonest symptomatic expressions of MDBA in adults (in Gautier's survey, neuralgia of fifth cranial nerve is present in 27 out of 204 cases), and it can be attributed to compression of the root of the fifth cranial nerve at the level of the pons. Furthermore, the symptoms of the pain attacks do not appear to be compatible with the criteria proposed by the International Headache Society Classification [18].

From a pathogenic point of view, we believe that a correlation may exist between attacks of pain (both migrainelike and ice-pick-like) and the vascular abnormality. This hypothesis is supported by the neuroradiological images obtained from both patients which show that the dolichobasilar artery is contiguous with the trigeminal root.

The strictly left-sided location of the painful attacks in both patients is particularly interesting and in accordance with the findings of Harsh et al. [15] which suggest that anomalies of the vertebrobasilar system are mainly localized on the left side. Left predominance may reflect the commonly occurring asymmetry in the diameter of the two vertebral arteries or a more efficient transmission of pulse pressure to the left vertebral artery from a subclavian artery that originates from the aortic arch rather than from the brachiocephalic trunk.

The association between trigeminal neuralgia and cluster headache has been described as cluster tic syndrome by Alberca and Ochoa [12], and in one of their patients, cluster tic syndrome coexisted with an ectatic basilar artery. In case 1, neuralgic attacks, similar to ice-pick-like pain [19], were present in association with migraine-like attacks. The presence of a vascular malformation compressing the trigeminal sensory nerve root would be sufficient to explain both types of pain attack. Indeed, the involvement of the medium-myelinated A-beta fibers may be responsible for the neuralgic pain [20], while an alteration of unmyelinated fibers is suggested to be involved in the pathogenesis of migraine [21]. Cruccu et al. [20] also suggest that the root entry zone of the trigeminal nerve may be a 'locus minoris resistentiae', since the myelin sheath provided by the Schwann cells is replaced by the oligodendroglia. It must be stressed that carbamazepine treatment led to remission of the pain.

Our study demonstrates the diagnostic effectiveness of noninvasive neuroradiology, such as MRI and MRA, and the need for a thorough study of cases with pain characteristics that seem similar to those of migraine, but have anomalous or atypical aspects.

Eur Neurol 1999;41:10-14

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