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Case Report

Collateral circulation through the vasocorona of the cervical spinal cord in a case of moyamoya syndrome

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Abstract. The spinal cord is covered by a functionally important pial arterial network described by Adamkiewicz in 1881 under the name of vasocorona. This arterial network loosely interconnects the anterior and posterior longitudinal axes and may become enlarged under pathological circumstances. We report here a case of moyamoya syndrome in a 2-year-old boy with bilateral vertebral artery occlusion, in which the collateral supply to the basilar artery occurred via a dilated portion of the vasocorona of the upper cervical spinal cord.

Keywords: Vasocorona, moyamoya, Keutel syndrome

1. Introduction

A 2-year-old boy with Keutel syndrome presented with an episode of right hand shaking. Magnetic resonance imaging and magnetic resonance angiography documented bilateral internal carotid artery stenosis. In addition, a markedly increased vascularity was observed around the upper cervical spinal cord, of unclear significance (Figs 1A and 1B). Diagnostic cerebral digital subtraction angiography showed bilateral carotid and vertebral artery occlusions, the latter at the level of the V4 segment. Collateral supply to the basilar artery was provided by the left costocervical trunk via an enlarged anterior spinal artery (Fig 2A), and by the right vertebral artery through a prominent arterial network developed from the upper cervical vasocorona (Figs 2B-2D). Figure 3 illustrates the upper cervical vasocorona, acting as a collateral pathway for the posterior fossa arterial vascularization.

2. Discussion

The earliest arterial vascularization of the spinal cord is derived from the spinal branches of the primitive intersegmental arteries [1,2]. The radicular arteries accompanying each spinal nerve root provide, as they reach the surface of the cord, a rich arterial anastomotic network called the primitive pial arterial plexus. The selection of preferential paths within this network later determines the formation of several longitudinal arterial chains, including the anterior spinal artery and the posterior-medial and

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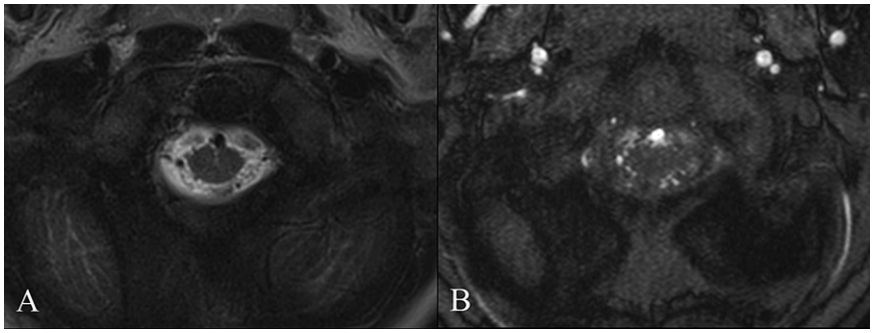

Fig. 1. A 2-year-old boy with Keutel syndrome and bilateral vertebral artery occlusions. (A) Magnetic resonance imaging, T2-weighted image, axial plane, showing prominent perimedullary flow voids indicating the presence of abnormal vascular structures at the level of the upper cervical spinal cord. (B) Magnetic resonance imaging, time-of-flight magnetic resonance angiography, axial plane, confirming the upper cervical hypervascularity suggested by the T2-weighted sequence.

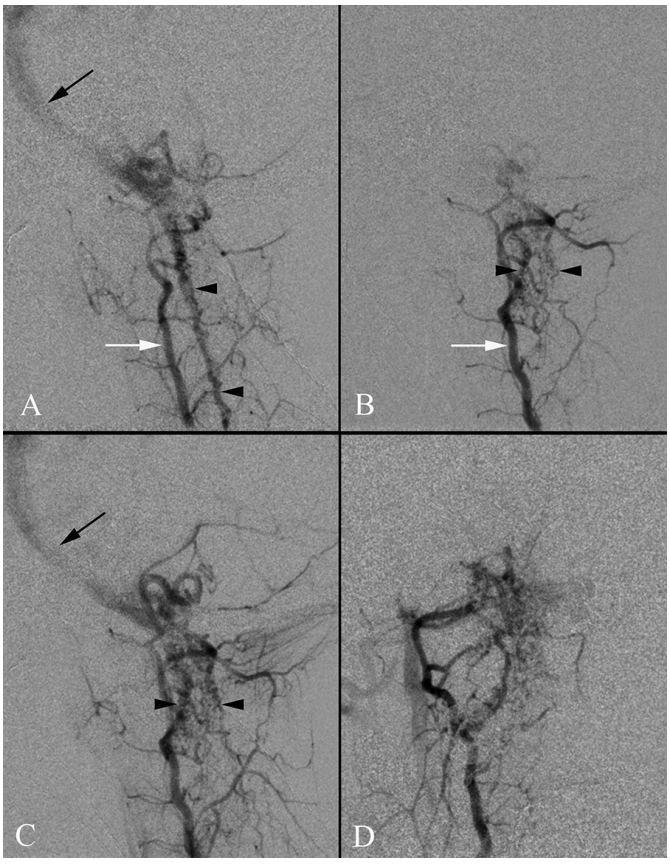

Fig. 2. (A) Digital subtraction angiography (DSA), left subclavian artery injection, lateral projection. The left vertebral artery (white arrow) is occluded at the level of its V4 segment. The basilar artery (black arrow) is principally opacified via a prominent anterior spinal artery (arrowheads) originating from the left costocervical trunk. (B) DSA, right vertebral artery injection, lateral projection. In this early arterial phase, the vertebral artery (white arrow, occluded at the level of its V4 segment) is seen supplying a diffuse arterial network surrounding the upper cervical spinal cord (arrowheads). (C) DSA, right vertebral artery injection, lateral projection. In the late arterial phase, the distal vertebral artery, part of the right posterior inferior cerebellar artery, and the basilar artery (black arrow) are opacified via the blood transiting through the dilated cervical perimedullary network (vasocorona) (arrowheads). (D) DSA, right vertebral artery injection, postero-anterior projection. This view better delineates the multiple V3 and V4 branches supplying the dilated upper cervical vasocorona at C1, C2, and C3.

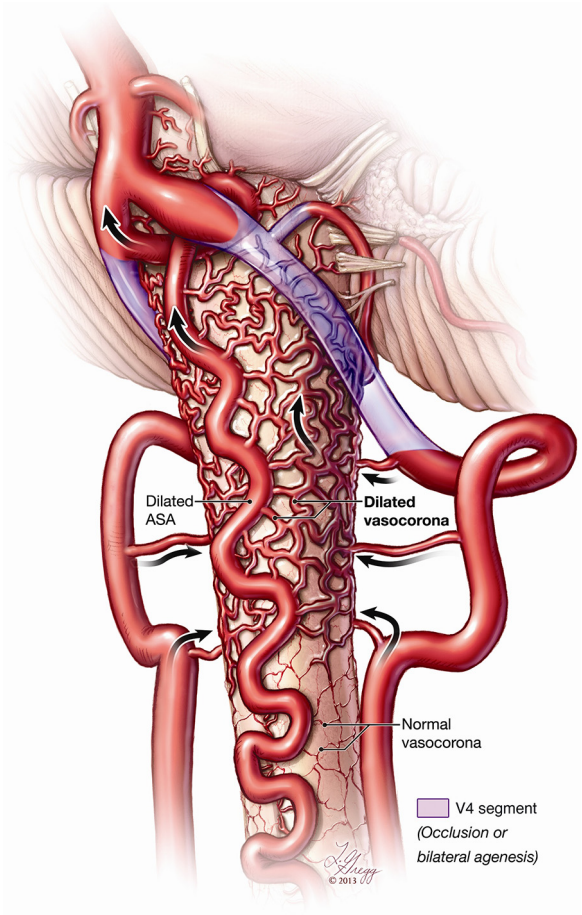

Fig. 3. Artistic representation of the vascular configuration seen in our patient. The occluded V4 segments are indicated by purple shading. The prominent dilatation of the upper cervical vasocorona acts as a collateral pathway for the posterior fossa arterial vascularization.

posterior-lateral spinal arteries [3,4]. The remainder of the primitive pial plexus takes, at the adult stage, the form of an arterial network stretched over the surface of the cord, loosely interconnecting the anterior and posterior longitudinal axes, the vasocorona [1].

While portions of the vasocorona may be enlarged under pathological circumstances, in the presence of a vascular malformation for example, it cannot be documented in its normal state by modern noninvasive imaging techniques or even by digital subtraction angiography. Collateral pathways involving the anterior or posterior spinal chains have been well described, notably in association with coarctation of the aorta [5] at times leading to a spinal steal syndrome [6]. On the other hand, the potential role of the vasocorona as a collateral pathway has, to our knowledge, not been previously reported. It is unclear what type of effect this abnormal utilization of the vasocorona might have on the underlying spinal parenchyma. Transient cord dysfunction from a steal syndrome or, after a longer evolution, permanent ischemic damage could be potential complications of this particular anatomy. From a pragmatic standpoint, the increased perimedullary vascularity associated with collateralization through an engorged portion of the vasocorona has a nonspecific appearance on magnetic resonance imaging; it should therefore be included in the differential diagnosis of spinal pial enhancement, along with conditions such as pial metastases or sarcoidosis for example [7].

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