JBR–BTR, 2011, 94: 293-294.

MRI FINDINGS IN GIANT PONTINE CAPILLARY TELANGIECTASIS ASSOCIATED WITH A DEVELOPMENTAL VENOUS ANOMALY

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We report a 32-year-old woman with an exceptionally large capillary telangiectasia in the brainstem which is associated with a developmental venous anomaly (DVA). Her major problems were nystagmus in both eyes, binocular diplopia, gait abnormalities, cerebellar ataxia, slightly disturbed finger-nose test, an instable Romberg test and obvious dysartria. The diagnosis was made on the basis of specific imaging findings, and the use of gradient echo-weighted images proved to be helpful in making a differential diagnostic decision.

Key-word: Telangiectasia.

Cerebral vascular malformations, which include capillary telangiectasias, developmental venous anomalies (DVA), cavernous angiomas and arteriovenous malformations are nonneoplastic developmental anomalies that present a variety of clinical patterns, ranging from asymptomatic to fatal intracranial hemorrhage. We report a case of an exceptionally large capillary telangiectasia in the brainstem, which is associated with a DVA. The use of gradient echo-weighted images proved to be helpful in making a differential diagnostic decision.

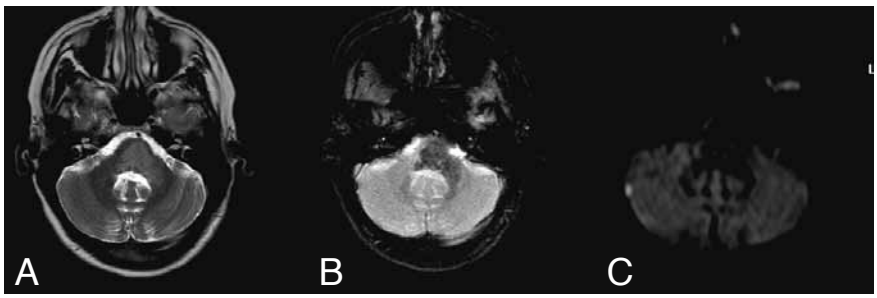

Fig. 1. — A. Axial T2-weighted image: a large hyperintense area is seen in the pons. B. Gradient echo-sequence: low signal intensity is present in the lesion. C. DWI (b = 1000): no diffusion restriction is seen.

Case report

A 32-year-old woman was admitted to our neurology department with complaints of visual disturbance, slurred speech and gait ataxia. Physical examination revealed nystagmus in both eyes, binocular diplopia, gait abnormalities, cerebellar ataxia, slightly disturbed finger-nose test, and instable Romberg test. An obvious dysartria was noted. An EEG was obtained and was negative. All laboratory findings were within normal limits. Magnetic resonance (MR) imaging of the brain was performed using standard spin-echo T1- weighted and T2-weighted, gradient-echo sequences and diffusion-weighted images. After intravenous injection of gadolinium axial and sagittal T1-weighted images were made. The T2-weighted images (Fig. 1A) demonstrated a large region with increased signal intensity extending in the midpons and lower pons, and in the upper medulla oblongata. The lesion was hardly visible on the unenhanced axial T1-weighted images (Fig. 2A), and showed a low signal intensity on the gradient echo-weighted sequence (Fig. 1B). On the diffusion-weighted images (DWI) (b = 1000) no restriction was noted (Fig. 1C). No mass effect was present. After intravenous gadolinium injection diffuse and moderate enhancement was seen on the axial and sagittal T1-weighted images (Fig. 2B, 2C), and the borders of the enhancing region were brush-like. Within the moderately enhancing region an enlarged stronger enhancing central vessel extended to the pial surface. In view of the specific location of the abnormal findings, the absence of mass effect, and with the finding of a larger vessel running through this midbrain portion to the pial region the diagnosis of combined presence of capillary telangiectasia and DVA could be made. In this patient a wait-and-see policy was conducted.

Discussion

The true incidence of capillary malformations or telangiectasias of the brain is difficult to discern because most are likely to be clinically asymptomatic. Estimates from autopsy series suggest they are not uncommon, representing approximately 16% to 20% of all CNS vascular malformations (1). Capillary malformations represent histologically benign collections of dilated capillaries interposed within normal brain parenchyma (2).

The area of involvement of the brain is typically small, ranging from several millimetres to 2 centimeters in size (3, 4). With a craniocaudal extention of almost 4 cm the lesion in our case is uncommonly large. Sayama CM et al. (5) reported, brain capillary telangiectasias can cause symptoms, that may actually be related to the size effect of the lesion. When symptoms occur, they are most likely due to the associated vascular malformations, although occasional capillary telangiectasias alone may be symptomatic. Hemorrhage seen in association with capillary telangiectasia almost always arises from an associated vascular malformation and only rarely from the capillary telangiectasia (6, 7).

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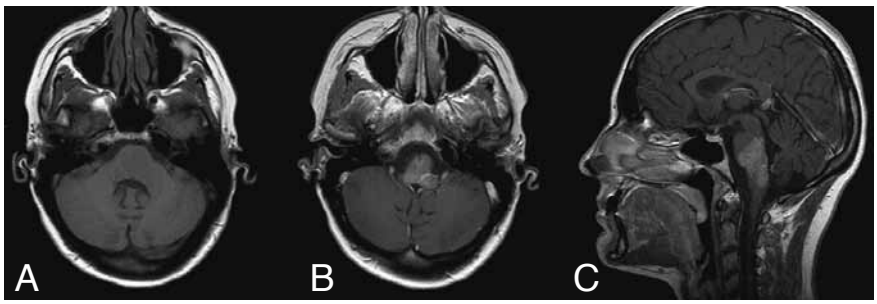

Fig. 2. — A. Axial T1-weighted image: the lesion is hardly visible. B. Axial enhanced T1-weighted image: there is moderate enhancement of the lesion with brush-like borders. A strongly enhancing tubular structures crosses the lesion corresponding to a DVA. C. Sagittal enhanced T1-weighted image: Diffuse and moderate enhancement with a craniocaudal extension over 4 cm.

Common sites of involvement include the pons – as is the case in our patient –, but also cerebral hemispheres and spinal cord (2, 4). In one series of 18 presumed capillary telangiectasias, only two were found outside of the brain stem (temporal lobe and head of caudate nucleus) (3). Furthermore, a case of an unusually large, biopsy-confirmed capillary telangiectasia involving the basal ganglia was also reported by Castillo M et al. (6). Capillary telangiectasias and transitional lesions can be found at the periphery of cavernous angiomas in autopsy series. Because of these similarities, Rigamonti et al conclude that the two lesions represent "a spectrum within a single pathological entity" (8). Barr et al discuss that capillary telangiectasia and cavernous angiomas might represent the spectrum of changes that can occur as a result of venous restriction (3). McCormick et al. suggest that " elevated venous pressure in a venous angioma leads to ectatic dilated microvasculature, which represents an acquired telangiectasia that evolves toward a cavernous malformation" (9).

On the gradient echo-weighted images low signal intensities are seen in the region with capillary telangiectasis. This decrease in signal is explained by the presence of increased intravascular deoxyhemoglobine (3). This signal decrease is an interesting differential diagnostic finding, since it is not seen in case of a low grade glioma, which is the most frequently noted similar lesion in the brainstem. Moreover low grade gliomas do not enhance and show mass effect. We did not performe susceptibility-weighted imaging (SWI), but SWI was useful for imaging diagnosis as it demonstrated marked signal loss of the lesion, if the lesion did not show characteristic signal loss on conventional gradient-echo images (10).

Other differential diagnostic entities in this pontine location are demyelinating disease, infection, infarction or central pontine myelinolysis. In this patient demyelinating disease can almost for sure be excluded since no other abnormal regions were noted, especially not in a periventricular location, infection was improbable on a clinical basis, infarction was excluded on basis of the findings on the DWI. DWI seems to be a useful adjunct for the diagnosis of capillary telangiectasias which will facilitate the differential diagnosis concerning tumorous, inflammatory and ischemic lesions (11). Central pontine myelinolysis was excluded since the abnormal findings extent out of the pontine region and also on clinical basis (no history of alcohol abuse).

It cannot enough be emphasized that capillary telangiectatic lesions should not be biopsied because of the hemorrhage risk. In our patient a conservative observational policy was chosen.

In conclusion, this report shows that a very large pontine capillary telengiectasia associated with a venous angioma can present without hemorrhage, and illustrates that the use of gradient echo-weighted images increases the degree of confidence in a diagnosis that cannot be achieved with a biopsy.

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