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 dysarthria. 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.

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 extension 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. 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.
in the brainstem. Moreover low grade gliomas do not enhance and show mass effect. We did not perform 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, infarction 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 tumors, 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 telangiectasia 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.

References