Clinical signs associated with megadolichobasilar artery anomaly

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Abstract

The elongation and ectatic course of the basilar artery (BA), called megadolichobasilar artery anomaly (MDBAA) is a macroscopic description of a neuroradiologic finding. Clinically ischemic brain stem syndromes and peripheral cranial nerve disturbances especially of the trigeminal and facial nerves, cerebellar dysfunction and CSF circulation disturbances are observed. Seldom a subarachnoid hemorrhage is proven. In CT and MRI often a tumorlike mass with a ringlike contrast enhancement combined with a nonhomogeneous lesion due to partial thrombosis, is detected. Angiography in most cases shows fusiform dilatation of the BA, elongation of the top of the basilar trunk, asymmetric tortuosity or dislocation of the irregular shaped wall of the BA.

Fifteen patients within the last 12 years are demonstrated. The role of reticular fiber deficiency in the media and defects of the elastic lamina as the basis of these malformations is reported. We discuss coincidental findings of MDBAA with atherosclerosis, congenital factors causing generalized vasculopathy, metabolic disturbances in form of so called inborn errors of metabolism and endocrine deficiencies.

In cases with clinical signs the morbidity is remarkably high.

Keywords: Atherosclerosis, endocrine vasculopathy, inborn errors of metabolism, megadolichobasilar anomaly, pathogenesis.

1 Material and method

Fifteen patients with a proven MDBAA for the last 12 years are reported. In all cases CT, angiography and MRI in the last cases were performed. Arterial hypertension, smoking, diabetes mellitus, hyperlipaemia were present in 12 patients.

Two cases demonstrated metabolic disorders, one with a glycogenosis and another with a HGH secreting pituitary adenoma. Three patients developed an obstructive hydrocephalus with need of shunting. An intracerebellar hematoma was detected in one case. In another case SAH was observed, follow up angiography revealing a “growing BA” (Figures 3, 4, 5). A pronounced tortuosity with lateral dislocation of more than 20 mm was seen in three patients who suffered from trigeminal neuralgia. In these patients the asymmetric malformation reached the cerebello-pontine angle. Angiography revealed the ectasia and elongation of the BA, exceeding the sella more than 40 mm in one case (Figure 4).

In CT a space occupying enhancing lesion of the posterior fossa was found in 12 cases. A ringlike enhancement due to the vasa vasorum was seen in the wall of the partially thrombosed fusiform aneurysm (Figure 5). Clinically most often there was a combination of brain stem syndromes with cerebellar ataxia, pyramidal signs, cranial nerve dysfunction. In none of our cases hemifacial spasm was observed.

With MRI in sagittal planes the full extent of the malformation and it’s nature was well detectable.

2 Illustrative cases

Case I: 42-year-old male, hypertensive. Transient hemiparesis, hemihypesthesia, dysarthria, diplopia. CT (Figure 1) and angiography (Figure 2) demonstrating MDBAA with a diameter of 12 mm within the cerebellopontine angle, the tip of the BA 35 mm above the sella. Six days after the onset of symptoms a pontine hypodensity is well delineated.

Case II: Male, 42 years, hypertensive. Progressive cerebellar ataxia, dysarthria, deafness and facial weakness.
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the MDBAA in association with ectatic posterior cerebral arteries, carotid arteries and abdominal aorta.

**Case III:** Female, 77 years, recurrent HGH secreting pituitary adenoma with typical clinical signs of an acromegaly. Acute visual disturbances. MRI and CT demonstrating an intrasellar recurrence of a small adenoma, angiography (Figure 6) showing a MDBAA associated with fusiform ectatic carotid arteries and peripheral arteries.

3 Discussion

The incidence of MDBAA is differing in literature. Synonyms are giant aneurysm of BA (5/21/22), fusiform aneurysm [2, 4, 10, 11], ectatic or doli
dchoectatic BA [7] and tortuosity of BA [9, 13].

Müller [19] measured the length of the BA with 30–35 mm in the average. Yasargil [14] showed that the cranial bifurcation ends at the level of the posterior clinoid process in 50%. In more than 50% there is a straight course, in 40% there is a moderate lateral dislocation, due to hemodynamic factors by an asymetrically developed vertebral artery (Haverling [9]). Yu et al. [24] reviewed 10,000 angiograms and found 31 cases with MDBAA. Nijensohn et al. [20b] estimated the autopsy material at the Mayo clinic and found 20 cases within 20 years [20b].

Figure 1. CT demonstrating the MDBAA and a pontine hypodensity.

Angiography revealed a giant MDBAA with partial thrombosis. Months later onset of symptoms of an obstructive hydrocephalus requiring shunt procedure. One year later admission with clinical signs of shunt dysfunction whereas SAH was prooven. Controll angiography (Figures 3, 4) and CT (Figure 5) revealed an extensive “growing” of

Figure 2. Vertebralis angiography demonstrating the BA 35 mm above the sella and the tortuos course of the BA (same pat. as fig. 1 ~ Case 1).