Meningocerebral angiodysplasia

The uncommon simultaneous occurrence of an exuberant, angioma-like proliferation of superficial cerebral microvessels along with absence of the kidneys has been proposed to constitute a syndromic complex for which the term “meningocerebral angiodysplasia (or angiomatosis) with renal agenesis” (MCA-RA) is being descriptively used.

Case reports

2016

A girl with progressive left frontal tissue destruction starting at the age of almost 8 years. She manifested acutely with epileptic seizures accompanied by Broca aphasia as well as transient right hemiparesis. Due to refractory epilepsy developing over the next years, which originated from the left frontal lobe, the decision was made to proceed to epilepsy surgery. By then, her language functions had recovered despite progressive left frontal tissue-destruction, raising the possibility of a hemispheric shift of language. Clinical functional magnetic resonance imaging (fMRI) was conducted to localize brain regions involved in language production. A complex pattern of clear right-hemispheric dominance, but with some left-sided contribution was found. However, a Wada test suggested the left hemisphere to be critical, seemingly contradicting fMRI. Invasive electroencephalogram recordings could reconcile these results by identifying the fMRI-detected, residual left-sided activation as being relevant for speech production. Only by combining the localizing information from fMRI with the information obtained by two invasive procedures could the unusual pattern of late-onset language reorganization be uncovered. This allowed for extensive left frontal resection, with histology confirming meningocerebral angiodysplasia. Postoperatively, language functions were preserved and seizure outcome was excellent. The implications of the findings for presurgical assessments in children are discussed [1)].

2011

Rocha et al observed this constellation in one of a pair of dichorionic male twins following postpartal death in the 38th week of pregnancy. General autopsy revealed rudimentary metanephric anlagen made up of few residual glomeruli, cysts lined by flattened tubular epithelium, and islands of cartilage - corresponding to renal aplastic dysplasia. Largely inconspicuous with respect to its gyral pattern, as well as the configuration of the ventricular system, the brain microscopically showed extensive replacement of the cortex by a lattice of proliferating capillaries with necrosis of the intervening parenchyma. Minute foci of calcified necrosis were scattered in the deep subcortical white matter as well, while the ventricular ependyma and the subventricular germ cell layer remained remarkably intact. The cerebellum and brain stem appeared unaffected as well. Karyotyping of skin fibroblasts indicated a normal chromosome set of 46XY without gross structural anomalies. We interpret these findings as ones apt to being reasonably accommodated within the spectrum of MCA-RA. Although exceedingly rare, accurate identification of individual cases of MCA-RA is relevant both to differential diagnosis from its prognostically different look-alike “proliferative vasculopathy and hydranencephaly-hydrocephaly” (PVHH), and to refine the nosology of unconventional pediatric vascular malformations, for which the rather nonspecific label “angiodysgenetic necrotizing encephalopathy” is still commonly used [2)].

1986

A third case of meningocerebral angiodysplasia involving the cerebral cortex, with necrosis of both cortex and white matter is described in a neonate with Potter's syndrome. It is suggested that cortical vessel anomalies act as a local intracerebral shunt to produce periventricular infarction. This disorder differs from pure meningeal angiectasis without cerebral infarction and from the classic arteriovenous malformation of the central nervous system with massive systemic shunt, cardiomegaly, and neonatal cardiac failure. Meningocerebral angiodysplasia and renal agenesis appear to form a rare but distinct association [3].

1)

Lorenzen A, Wilke M, Alber M, Milian M, Bornemann A, Ernemann U, Rona S. Multimodal Assessment Reveals Late-Onset Hemispheric Shift of Language in a Child with Meningocerebral Dysplasia. Neuropediatrics. 2016 Jul 27. [Epub ahead of print] PubMed PMID: 27462834.

2)

Rocha C, Schäfer S, Kappeler A, Baumann M, Surbek D, Vajtai I. Meningocerebral angiodysplasia with metanephric induction failure: Broadening the spectrum of an emerging maldevelopmental syndrome. Pathol Res Pract. 2011 Jul 15;207(7):456-61. doi: 10.1016/j.prp.2011.05.001. Epub 2011 Jun 12. PubMed PMID: 21664057.

3)

Valdivieso EM, Scholtz CL. Diffuse meningocerebral angiodysplasia and renal agenesis: a case report. Pediatr Pathol. 1986;6(1):119-26. PubMed PMID: 3822929.