Primary Familial Brain Calcification

Pericytes are perivascular cells along capillaries that are critical for the development of a functional vascular bed in the central nervous system and other organs. Pericyte functions in the adult brain are less well understood. Pericytes have been suggested to mediate functional hyperemia at the capillary level, regulate the blood-brain barrier and to give rise to scar tissue after spinal cord injury. Furthermore, pericyte loss has been suggested to precede cognitive decline in mouse models of Alzheimer's disease. Despite this observation, there is no convincing causality between pericyte loss and the pathogenesis of Alzheimer's disease. However, recent loss-of-function mutations in PDGFB and PDGFRB genes have implicated pericytes as the principle cell type affected in primary familiar brain calcification (PFBC), a neuropsychiatric disorder with dominant inheritance.

Zarb et al., reviewed the role of the PDGFB/PDGFRB signaling pathway in pericyte development and briefly discussed homeostatic functions of pericytes in the brain. They provided an overview of recent studies with mouse models of PFBC and discuss suggested pathogenic mechanisms for PFBC with special reference to pericytes [1)].

Unclassified

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