FLNA (filamin A) and ZYX (zyxin) proteins were significantly higher in Moyamoya Disease serum compared with those in health controls (Log2FC >2.9 and >2.8, respectively). Immunofluorescence revealed an intimal hyperplasia in the superficial temporal artery and middle cerebral artery specimens of MMD. FLNA and ZYX proteins increased the proportion of endothelial cells in the S phase and promoted their proliferation, angiogenesis, and cytoskeleton enlargement. Mechanistic studies revealed that AKT (serine/threonine kinase)/GSK-3β (glycogen synthase kinase 3β)/β-catenin signaling pathway plays a major role in these FLNA- and ZYX-induced changes in endothelial cells.

This study provides proteomic data on a large sample size of MMD. The differential expression of FLNA and ZYX in patients with MMD and following in vitro experiments suggest that these upregulated proteins are related to the pathology of cerebrovascular intimal hyperplasia in MMD and are involved in Moyamoya Disease pathogenesis, with diagnostic and therapeutic ramifications [1].

Magaki et al. review the clinical and pathologic features of HPA and characterize the inclusions and brain tissue in which they are seen in surgical resection specimens from five patients with intractable epilepsy and HPA compared to five patients with intractable epilepsy without HPA using immunohistochemistry for filamin A, previously shown to label these inclusions, and a variety of astrocytic markers including aldehyde dehydrogenase 1 family member L1 (ALDH1L1), SRY-Box Transcription Factor 9 (SOX9), and glutamate transporter 1/excitatory amino acid transporter 2 (GLT-1/EAAT2) proteins. The inclusions were positive for ALDH1L1 with increased ALDH1L1 expression in areas of gliosis. SOX9 was also positive in the inclusions, although to a lesser intensity than the astrocyte nuclei. Filamin A labeled the inclusions but also labeled reactive astrocytes in a subset of patients. The immunoreactivity of the inclusions for various astrocytic markers and filamin A as well as the positivity of filamin A in reactive astrocytes raise the possibility that these astrocytic inclusions may be the result of an uncommon reactive or degenerative phenomenon [2]

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2)
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