Cerebral Amyloid Angiopathy–Related Inflammation in Down Syndrome–Related Alzheimer Disease

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A 59 years-old woman with Down syndrome (DS) and prodromal Alzheimer’s disease (AD) presenting progressive cognitive impairment in the previous year, with no focal symptoms, headache, seizures or accelerated cognitive decline, underwent 3T brain MRI showing cortico-subcortical tumefactive FLAIR hyperintensities, associated with microbleeds and superficial siderosis (Figure 1). These FLAIR abnormalities have moved spatially compared to a previous MRI, suggesting an inflammatory process (Figure 2). The diagnosis of cerebral amyloid angiopathy-related inflammation (CAA-ri) was made. DS is a genetic form of AD and has increased CAA prevalence. As future anti-amyloid trials will likely include individuals with Down syndrome, neurologists must be aware of CAA-ri in this population.

References:


Figure 1. Spatial relationship between microbleeds, superficial siderosis and FLAIR abnormalities.

Baseline MRI shows multiple microbleeds in the temporo-occipital regions (A) and cerebellum (B) and superficial siderosis in the right occipital lobe (red arrows in C). (D) and (E) show the anatomic relationship between inflammatory changes on FLAIR (E) and microbleeds on SWI (D). FLAIR: fluid-attenuated inversion recovery; SWI: susceptibility-weighted imaging.
Figure 2. Changes in the location of inflammatory abnormalities.

Changes in the location of FLAIR hyperintensities between the current and the previous MRI (2 years before). Red arrows show inflammatory changes present in the previous and absent in the current MRI. Blue arrows show inflammatory changes absent in the previous and present in the current scan. FLAIR: fluid-attenuated inversion recovery.
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