



**Online Resource 1** Neuropathology of definite cerebral amyloid angiopathy (CAA) cases. a) Leptomeningeal and penetrating cortical arteries with thickened Congo red-positive walls, demonstrating apple-green birefringence under the polarized microscope (insets). b) Immunostaining for amyloid- $\beta$  reveals denegerated vessel walls with typical double-barrel formation, with occasional widening of the perivascular space and perivascular amyloid- $\beta$  deposition (inset). c) Subcortical microbleeds with erythrocyte extravasation, hemosiderin deposition, and characteristic central pallor with a central arteriole/capillary (often occluded; lower-left inset (Crossmon)), occasionally with immunopositivity for amyloid- $\beta$  (upper-right inset). d) Cortical superficial (i.e., subarachnoid) siderosis with apparent siderophage activity (occasionally also laden with diffuse amyloid- $\beta$ -immunopositive material; inset). e) All 3 patients with definite CAA demonstrated different amount of amyloid- $\beta$  plaque pathology in addition to CAA. f) Only 2 of the 3 definite CAA patients showed Tau pathology in the examined regions with neurofibrillary tangles and dystrophic neurites (inset), reflecting the ~70% overlap with Alzheimer's disease pathology.