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LETTER TO THE EDITOR

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Commentary to “A 44-year-old female with familial Mediterranean fever, cardiomyopathy and end stage renal disease” by Magaki et al.

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We would like to comment on the “Case of the Month” by Magaki et al, entitled “A 44-year-old female with familial Mediterranean fever, cardiomyopathy and end stage renal disease” (3). In this case report, the authors describe the autopsy findings in a patient with systemic AA amyloidosis secondary to familial Mediterranean fever. The patient also had cardiomyopathy with severely reduced left ventricular ejection and death due to cardiac arrest; however, no details about cardiovascular risk factors, cardiac pre-mortem explorations and heart gross examination were provided. Amyloid was found in vessels of the plexus choroid and pituitary gland, and was stained with anti-AA antibody. In accordance with the systemic distribution of AA amyloidosis, amyloid was also detected in the heart, kidneys, ovaries, thyroid, and spleen. This presentation, along with the title of the case report, indicates that the authors believe that cardiomyopathy was a manifestation of systemic AA and should have been viewed by the readers of *Brain Pathology* as a clue for the final diagnosis. We believe that insinuating a causal relationship between AA amyloidosis and cardiomyopathy is extremely questionable. Indeed, it is important to keep in mind that cardiac involvement is generally not a clinically relevant feature during AA amyloidosis (1), in opposition with AL and ATTR amyloidoses. Despite the absence of detailed cardiac histology in the case report and the scarcity of literature on this point, it is likely that,

similarly to what was found in the plexus choroid, distribution of amyloid was limited to vessel walls of cardiac microcirculation, without involvement of myocardial interstitium. Conversely, in AL and ATTR amyloidoses, amyloid is predominantly interstitial (2), which probably explains why these two forms impair cardiomyocyte function and electric conduction, whereas AA amyloidosis does not. In summary, we believe that cardiomyopathy, a rather frequent condition with numerous possible causes, was fortuitously associated with clinically silent amyloid deposits in this patient’s case, and, more widely, should not be viewed as a feature of AA amyloidosis.

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