

Eosinophilic angiocentric fibrosis

Eosinophilic angiocentric fibrosis is a rare pseudotumorous disease of the orbit, upper and, exceptionally, the lower respiratory tract. It is probably one of the forms IgG4 associated disease, although the histological picture is somewhat different. Histopathologically, marked "bulbous" fibrotization (concentric layers of connective tissue) around small arteries is evident. The inflammatory infiltrate is rich in eosinophils. It is essential for histopathological diagnosis that necrosis, huge multinucleated cells or obvious manifestations of systemic vasculitis never occur.

Due to the rarity of the disease, the published generalizations are less reliable, because only individual cases or a series of a few cases can be analyzed. For example, Deshpande et al. analyzed 5 cases. In all patients they showed elevation of IgG4 in the serum, on immunochemical examination they showed IgG4 positive cells in 4 patients.

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