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Many faces of cutaneous vasculitis in eosinophilic granulomatosis with polyangiitis: An excellent model for histopathology of cutaneous vasculitides

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Introduction. – The wide clinical and histopathological spectrum of cutaneous vasculitis in EGPA has not been well described.

Methods. – Thirteen skin biopsy specimens obtained from seven EGPA cases with histopathologically proven necrotizing vasculitis were selected for clinical and histopathological study.

Results. – Three different cellular infiltrates of necrotizing vasculitis including neutrophilic (leukocytoclastic), eosinophilic and granulomatous vasculitis were found in 4, 3 and 3 cases, respectively. Vasculitis of different vessel size ranging from dermal small vessels (venules most often, arterioles less common) to subcutaneous muscular arteries and/or veins could coexist in the same patients or even at the same tissue specimen. Leukocytoclastic vasculitis with a mixed infiltrate of eosinophils was found in three cases with negative ANCA and no renal involvement, while an exclusively infiltrate of neutrophils was found in the remaining one with positive ANCA and renal involvement. Eosinophilic vasculitis was found in both dermal small vessels and subcutaneous arteries/veins, while histiocytic vasculitis was only found in subcutaneous arteries and veins. Small vessel vasculitis often presented as palpable purpura, while arteritis/phlebitis showed nodular erythema, livedo racemosa and subcutaneous nodules. Morphologic evolution of arteritis started as eosinophilic vasculitis followed by granulomatous vasculitis and ended at healed stage.

Discussion. – Both dermal small vessel vasculitis and subcutaneous muscular vessel vasculitis could be found on different occasions with different cutaneous manifestations. Dermal small vessel vasculitis showed two distinct phenotypes as eosinophilic vasculitis with negative ANCA at one end to neutrophilic vasculitis with positive ANCA at the other end.

Conclusion. – Different levels of vasculitis with different skin manifestations account for their diverse cutaneous manifestations in EGPA and provide an excellent model for learning histopathology of cutaneous vasculitides.

Further readings


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