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Cerebrovascular Disease Associated With Systemic Sclerosis: Case Series And Systematic Review

Background and aims: Central nervous system involvement in systemic sclerosis (SSc) has traditionally been considered uncommon. Recent studies suggest that SSc might be associated with an increased risk of cerebrovascular disease (CBVD), independent of conventional cardiovascular risk factors. We present the largest case series of symptomatic CBVD in SSc patients and the first systematic literature review of SCL and CBVD association.
Methods: A retrospective chart review of patients with SSc and possible CBVD seen at Henry Ford (1980-1997), Wayne State University (1998-2001), Mount Sinai (2001-2009), SUNY Downstate University (2010-2021) was performed.
MEDLINE/EMBASE/WoS were searched for “systemic sclerosis”, “scleroderma”, “CREST”, “cerebrovascular”, “cerebrovascular accident”, “cerebrovascular disease”, “stroke”.
Results: 14 patients with SSc and CBVD were included in our case series (mean age 48.5 years, 85% female). CBVDs were ischemic stroke (n=9, 64.1%), hemorrhagic stroke (n=1, 7.2%), cerebral venous thrombosis (n=1, 7.2%), ischemic optic neuropathy (n=1, 7.2%), possible ischemic stroke (negative CT head) (n=2, 14.3%).
Systematic review: 932 abstracts were screened after removal of duplicates; 110 articles were selected yielding 93 patients (mean age 48 years, 78.5 % female). CBVDs were vasculitis (n=22, 23.7 %), any intracranial hemorrhage (n=19, 20.4%), ischemic stroke (n=18, 19.3%), unruptured aneurysms (n=10, 10.7%), small vessel disease (n=6, 6.4%), Moya-Moya syndrome (n=4, 4.3%), others (n=14, 15%).
Conclusions: We identify a heterogenous spectrum of CBVD etiology in SSc patients, with ischemic stroke being the most common in our case series, and vasculitis in literature. A complex interaction between chronic inflammation, autoimmune mechanisms, and endothelial dysfunction seems to underlie the heterogeneity of CBVD in SSc.