Abstract #: 06

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Title: Giant Cell Arteritis in a Patient with Limited Systemic Sclerosis Treated with Tocilizumab: A Case Report

Objective(s): The co-existence of systemic sclerosis (SSc) and a primary vasculitis is rare, with most reported cases consisting of ANCA-associated vasculitis. Overlap of SSc and giant cell arteritis (GCA) has rarely been reported, with only six cases thus far published in the English literature. We wish to report the first case of SSc-GCA overlap to be treated with tocilizumab.

Method(s): Retrospective chart review of clinical presentation, diagnostic elements, and evolution on treatment of a patient with SSc-GCA overlap.

Result(s): A 75-year-old woman with a 3-year history of limited anti-centromere-positive SSc presented with 6-weeks of bitemporal headaches and jaw claudication. She had no new visual symptoms, but an ophthalmologic exam confirmed left anterior ischemic optic neuropathy and probable early right side involvement. Inflammatory markers were elevated (C-reactive protein 33.2 mmol/L, erythrocyte sedimentation rate 28 mm/hr). Anti-neutrophil cytoplasmic antibodies were negative. Doppler ultrasonography was positive for halo and compression signs. Biopsy of the left temporal artery showed an active lymphohistiocytic arteritis. No fibrinoid necrosis or periadventitial vessel involvement were found to suggest an alternative diagnosis such as ANCA-associated vasculitis. PET scan showed no signs of large vessel vasculitis or neoplasia. Given the potential risk for scleroderma renal crisis, intermediate doses of intravenous methylprednisolone and oral prednisone were given, and tocilizumab was added as a steroid-sparing agent for a total period of two years. Her GCA relapsed 8 months after steroid cessation while on tocilizumab, with recurrence of bitemporal headaches, scalp tenderness and new musical tinnitus, which resolved with glucocorticoid resumption. Her GCA relapsed once again 6 months after tocilizumab cessation, with new large vessel vasculitis on PET scan, halo and compression signs on doppler ultrasonography, and high inflammatory markers requiring resumption of prednisone and tocilizumab.

Conclusion(s): We report the first case of GCA and SSc overlap to be treated with tocilizumab. Only six other cases of GCA and SSc overlap have been reported since 1978, and all were treated with glucocorticoids alone. Given the potential risk of glucocorticoids exposure in SSc patients, tocilizumab may be considered as a steroid-sparing agent in the context of overlapping GCA. Despite its rarity and lack of a clear pathophysiologic link between the two entities, GCA should be added to the differential diagnosis of headaches in SSc, given its potentially serious consequences.