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Clinical spectrum and outcomes of idiopathic inflammatory myopathies at Chris Hani Baragwanath Academic Hospital

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Abstract

Background: Idiopathic inflammatory myopathies (IIM) are rare diseases for which there is a paucity of data in Africa. We undertook a retrospective records review of clinical and laboratory features of patients with IIM attending a tertiary service in Gauteng, South Africa.

Patients and methods: Case records of patients seen between January 1990 and December 2019 and fulfilling the Bohan and Peter criteria for IIM were reviewed for demographics, clinical features, special investigations and drug therapy.

Results: Of 94 patients included in the study, 65 (69,1%) had dermatomyositis (DM) and 29 (30,9%) had polymyositis (PM). Overall, the mean (SD) age at presentation and disease duration were 41,5 (13,6) and 5,9 (6,2) years, respectively. 88 (93,6%) were Black Africans. The most common cutaneous features in DM patients were Gottron's lesions (72,3%) and abnormal cuticular overgrowth (67,7%). Dysphagia was the most common extra-muscular feature (31,9%), more so in PM than DM (p=0,02). Creatine kinase, total leucocyte count and CRP were similarly higher in PM than DM patients (p=0,006, 0,002, 0,01, respectively). Anti-nuclear and anti-Jo-1 antibodies were positive in 62,2% and 20,4% of patients tested, respectively, the latter significantly more in PM than DM patients (OR=5.1, p=0,03) and more likely to be positive with ILD (p=0,001). Corticosteroids were prescribed in all patients, 89,4% had additional immunosuppressive drugs and 6,4% required intensive/high care. Malignancies occurred in three patients, all of whom had DM. There were seven known deaths.

Conclusion: The present study provides further insights into the spectrum of clinical features of IIM, especially cutaneous features of DM, anti-Jo-1 antibodies and associated ILD, in a cohort of predominantly black African patients.