A case of indolent systemic mastocytosis with extensive cutaneous involvement

WAON Wickramasinghe¹, N Ediriwickrama¹

A 46 year old man presented with progressive cutaneous mastocytosis. He had no fever, gastrointestinal symptoms, organomegaly, cytopenia, skeletal involvement or liver derangement. Bone marrow biopsy revealed low mast cell burden with focal infiltrates of circumscribed lesions with central core of lymphocytes surrounded by aggregates (>15 cells/cluster) of mast cells, of which >40% are abnormal and spindle shaped, admixed with eosinophil precursors (Figure 1 and 2). Focal fibrosis was noted with no evidence of dysplasia, myeloproliferation or any other haematological malignancy. Based on WHO classification of tumours of haemopoietic and lymphoid tissue the features were consistent with Indolent systemic mastocytosis.

Figure 1. Bone marrow trephine biopsy (H&E stain x 10) showing focal infiltrates of circumscribed lesions.

¹Consultant Haematologist, Department of Haematology, Teaching Hospital, Kurunegala, Sri Lanka.

Correspondence: Dr. Omega Wickramasinghe
E-mail: omeewaits@yahoo.com
Figure 2. Bone marrow trephine (H&E stain x 40) high power view of circumscribed lesion with a central core of lymphocytes surrounded by aggregates of mast cells.

Figure 3. Bone marrow trephine biopsy (IHCX10) CD 117 demonstrated 20-25% marrow infiltration by mast cells.

References