Non Cutaneous, Non Langerhans Cell Histiocytoses: A Diagnostic Dilemma

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54 year old female presented with multiple lytic bony lesions along with B symptoms. Haemogram done showed mild anemia along with eosinophilia. Bone marrow examination was done considering possibility of multiple myeloma/metastatic infiltration. Aspirate smears were cellular and showed atypical cells. These cells were polygonal to elongated with basophilic cytoplasm, oval to elongated nucleus, fine chromatin and inconspicuous nucleoli (Fig. 1a, b). Bone marrow biopsy showed multiple foci of infiltration by these cells (Fig. 1c, d). The cells were strongly positive for CD68, CD163, Vimentin and S-100

and negative for CD21, CD23, CD56, CD1a, CK and CD45 (Fig. 2). Langerin IHC done from referral laboratory was negative. Test for acid fast bacilli and lepra bacilli were negative. In view of cytomorphological features, absence of cutaneous lesions along with very strong CD68 and CD163, a final diagnosis of ‘Miscellaneous Non-Cutaneous Non-Langerhan cell Histiocytoses’ was made [1, 2]. This is a newly described category in 2016 revised classification of histiocytic society and is being reported because of its rarity. The patient was started on interferon therapy 3 million units twice weekly and is currently (5 month follow up) doing well without fever and malaise along with normalization of weight.

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Fig. 1 Giemsa stained smears showing oval to elongated pleomorphic cells (Giemsa, A = 200 $\times$, B = 400 $\times$). The same cells seen forming granulomatous lesions in marrow biopsy (H&E, C = 200 $\times$, D = 400 $\times$)

Fig. 2 Immunohistochemistry on bone marrow biopsy section showing strong CD68, CD163, S100 and Vimentin (200 $\times$ magnification)

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Compliance with Ethical Standards

Conflict of interest All authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with human participants performed by any of the authors.

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