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Multisystem Langerhans cell histiocytosis: A diagnostic chameleon

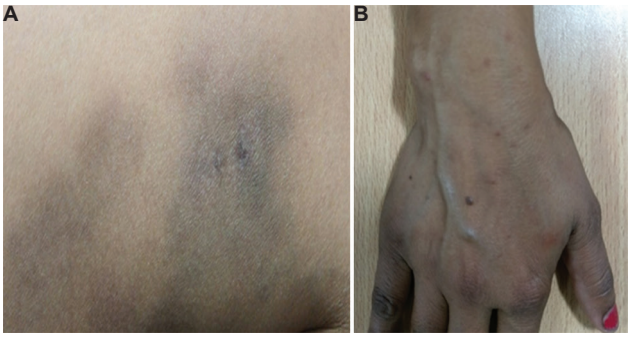

Fig. 1. (**A**) Hyperpigmented ill-defined macular rash on medial aspect of the right thigh. (**B**) Petechial rashes on the hand and forearm.

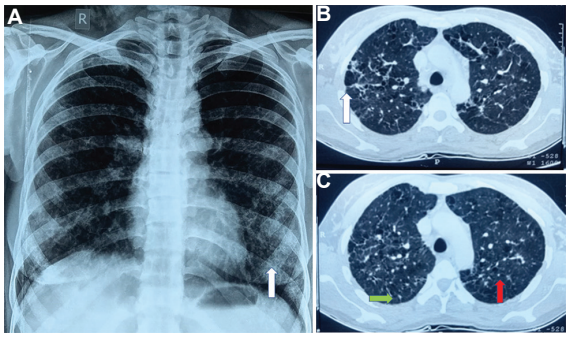

Fig. 2. (**A**) X-ray chest posteroanterior view showing bilateral reticular opacity (white arrow). (**B** & **C**) High-resolution computed tomography chest lung window showing septal thickening (green arrow), randomly distributed bizarre cysts (white arrow) and multiple centrilobular nodules (red arrow).

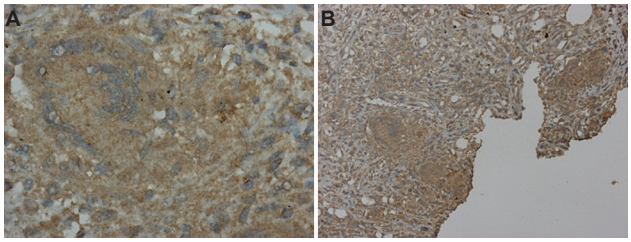

Fig. 3. (**A**) Photomicrograph of skin biopsy (×10); positive for CD1a. (**B**) Photomicrograph of skin biopsy showing epithelioid cell granuloma positive for CD1a (×40).

A 32 yr old non-smoker female† reported to the department of Dermatology, ESI-PGIMSR (Employee State Insurance-Postgraduate Institute of Medical Sciences & Research), New Delhi, India, in November 2018, for remitting and relapsing rashes all over the body for the last two years (Fig. 1A and B) with low-grade fever, malaise and weight loss. Routine haematological investigations were normal. The skin biopsy of the thigh region showed epithelioid cell granuloma with necrosis. The patient was started on anti-tubercular therapy. She was referred to the Pulmonary Medicine department for cough and abnormal chest radiograph (Fig. 2A) and the high-resolution computed tomography (HRCT) of the chest (Fig. 2B and C). The sputum was negative for acid-fast bacilli. The skin biopsy of the patient was re-evaluated as the radiological findings were not indicative of tuberculosis. On immunohistochemical staining, it was positive for S-100 and CD1a (Fig. 3A and B) confirming the diagnosis of systemic

†Patient's consent obtained to publish clinical information and images.

Langerhans cell histiocytosis. The patient had near-normal lung function on spirometry with 48 per cent diffusion capacity of carbon monoxide. At six months of follow up, the forced vital capacity dropped by 200 ml, but still was 77 per cent predicted. The patient is continued on oral prednisolone of 5 mg/day, with her symptoms controlled.

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***Conflicts of Interest*:** None.

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