Cluster of Differentiation 68 Negative Eruptive Adult Xanthogranuloma

Amrithaa Muralitharan1, Reena Rai2, Umamaheswari Gurusamy3

1Postgraduate; 2Professor and HOD, Department of Dermatology; 3Professor, Department of Pathology, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India.

Xanthogranuloma is a benign, asymptomatic, self-resolving disease of non-Langerhans cell histiocytosis (LCH). Herein, we report a case of multiple cutaneous eruptive xanthogranulomas with hypertriglyceridemia and central nervous system (CNS) manifestation in a young adult. A 28-year-old male presented with multiple, asymptomatic, skin-colored, and reddish–brown raised lesions over the face, neck, axillae, and forearms for 2 years. Cutaneous examination revealed multiple skin-colored, reddish–brown papules, nodules of varying sizes, few grouped, present all over the face, predominantly involving eyelids, periorbital area, nose, perioral area (Fig. 1A), neck, bilateral axillae (Fig. 1B), and bilateral cubital fossae which were nontender and soft. Few ulcerated nodules were seen in the axillae (Fig. 1B). The oral cavity, palms, and soles were normal. A differential diagnosis of eruptive xanthoma and xanthoma disseminatum was made. Routine blood investigations were within normal limits and lipid profile showed high triglyceride (300 mg/dL), borderline high total (208 mg/dL), and low-density lipoprotein (159 mg/dL) cholesterol levels. The ophthalmologic examination was normal. Histopathology revealed a dense diffuse infiltrate of cells comprising foamy macrophages, histiocytes, lymphocytes, plasma cells, neutrophils, and eosinophils in the dermis (Fig. 2A). Spindle cell proliferation with focal storiform pattern (Fig. 2B) and a few Touton type giant cells were observed (Fig. 2C). The foamy cells were negative for a cluster of differentiation 68 (CD68), melan-A, cytokeratin, and S-100 immunostains. Later, the patient developed weakness in the right upper and lower limbs and difficulty in speaking, for which contrast-enhanced magnetic resonance (MR) imaging with MR angiography of the Circle of Willis and neck was done, which revealed multiple focal lacunar infarcts at the midbrain and cerebellum on the left side. He was diagnosed with right hemiplegia with aphasia and posterior circulation stroke, possibly because of hypertriglyceridemia, and treated accordingly. Xanthogranuloma presents as red, yellow, or brown papules, nodules predominantly affecting infants, children, and rarely adults. The majority of the cases are solitary and multiple lesions occurring in an eruptive manner are rare. Xanthogranuloma is usually a normolipemic, non-LCH, but it has also been associated with hypertriglyceridemia, as seen in our case. Extra cutaneous involvement can also occur in the eye, CNS, lung, liver, and kidney mostly in the juvenile variant. A newly revised classification of “histiocytosis”
Correspondence

was devised, dividing them into five groups, consisting of L, C, M, R, and H groups. Adult xanthogranuloma is included in the C-group comprising non-LCH of the skin and mucosa. The non-LCH group is defined by the accumulation of macrophages/dendritic cells that do not meet the ultrastructural and phenotypic criteria for the diagnosis of Langerhans cells. The non-LCH histiocytes are positive for CD-68 and factor XIIIa and negative for CD-1a, and S-100.5 CD68 is an organelle-specific marker for lysosomes rather than a lineage-specific marker. The negative CD68 expression in our case might be attributable to scant lysosomes in the mononuclear cells.5 Eruptive xanthoma and xanthoma disseminatum were considered differential diagnoses. In eruptive xanthoma, yellowish papules occur on the extensor aspect of limbs, buttocks, and mucosal surfaces and are usually associated with diabetes mellitus and hyperchylomicronemia. In xanthoma, disseminatum papules, plaques, and nodules occur with oral mucosa involvement and diabetes insipidus.2,4 Our case is unique because of the CD68 negativity, eruptive manner of presentation, hypertriglyceridemia, and CNS manifestation.

Figs 1A to B: (A) Multiple skin-colored, reddish–brown papules and nodules over the face; (B) axillae shows multiple skin-colored, reddish–brown papules and few ulcerated nodules

Figs 2A to C: (A) Upper and mid dermis is expanded by a dense diffuse infiltrate of cells comprising of foamy macrophages (red arrows), histiocytes, lymphocytes, plasma cells, neutrophils, and eosinophils (H and E, 40x); (B) spindle cell proliferation with focal storiform pattern (H and E, 10x); (C) a Touton type giant cell (black arrow) is seen (H and E, 40x)
REFERENCES


