Multiple Vegetating Granulomatous Nodules in a 94-year-old Man

Yi-Chieh Lin  Chien-Ping Chiang

CASE REPORT

A 94-year-old male patient presented with multiple vegetating and granulomatous nodules on the right groin and gluteal region in the recent one year (Fig. 1A). He was robust before without any other systemic diseases except oral erosion developed one week before he visited our clinic. Tentative diagnosis included cutaneous fungal infection, cutaneous T cell lymphoma, and vegetating blistering disease. Bacterial and fungal culture showed no growth of microorganism. An incisional biopsy was performed in the gluteal region and histopathology showed epidermal hyperplasia, and intraepidermal eosinophilic abscesses (Fig. 2). Direct immunofluorescence (DIF) studies of the lesion revealed intraepidermal IgG and C3 deposition (Fig. 3). Indirect immunofluorescence revealed circulating antiepithelial cell surface IgG. The patient responded well to topical occlusive clobetasol treatment and those lesions regressed a lot after one week (Fig. 1B).

Fig. 1
(A) 5x5 cm enlarged vegetative ulcerative nodules over the gluteal region.
(B) The plaque responded to topical occlusive clobetasol treatment and regressed a lot after 1 week.

Fig. 2
Histopathology of the tumor lesion showed epidermal hyperplasia and papillomatosis. It is characterized by the intraepidermal eosinophilic microabscesses (H&E, original magnification x40).

Fig. 3
DIF of the tumor lesion revealed intercellular deposits of IgG antibodies (DIF, original magnification x400).
DIAGNOSIS: Pemphigus Vegetans, Hallopeau Type

DISCUSSION

Pemphigus vegetans was described as a variant of pemphigus vulgaris. The mean age of onset is 40-50 years, and this late onset is extraordinary in the English literature. The disease was first described by Neumann in 1876 characterized by extensive, coalescent bullae and papillomatous growths appeared on it (Neumann type). Another variant with annular pustules that eroded to form verrucous, cobblestoned plaques called Hallopeau type. The later is more benign than the aggressive, unremitting bullous disorders of Neumann type (the usual type of pemphigus vegetans). The diagnosis depends on the histological features of epidermal hyperplasia, papillomatosis, intraepidermal eosinophilic abscesses, and acantholysis. And DIF findings shows strong intercellular deposits of IgG and C3.

The etiopathogenesis is not clear. Antibodies directed against 130- and 85-kd polypeptides of pemphigus vulgaris antigens have been detected in the serum of patients with the hallopeau type. Pemphigus vegetans has also been associated with some drug exposure, most frequently captopril and penicillamine. Besides, intranasal heroine was reported. A hypothetical mechanism is that these drugs had thiol groups may induce acantholysis.

Pyoderma-pyostomatitis vegetans (PD-PSV) is a rare, benign, eosinophilic pustular and vegetating mucocutaneous disease with unknown pathogenesis. It had the same clinical and histopathological features with pemphigus vegetans. Immunofluorescence technique is useful to differentiate between the two entities.

There are no standard recommendations for treatment for late onset pemphigus vegetans. Systemic glucocorticoids have been the mainstay clinical treatment of pemphigus vegetans. The addition of other immunosuppressive agents, such as cyclophosphamide, azathioprine, intramuscular gold or etretinate, may improve remission rates. The course and prognosis of our late onset Hallopeau type patient is excellent and the vegetative plaques diminished after 1 week steroid occlusive therapy.

REFERENCES