Case 14
A 42-year-old Thai male from Bangkok
Chief complaint: A 4-month history of multiple itchy erythematous papules and plaques on trunk

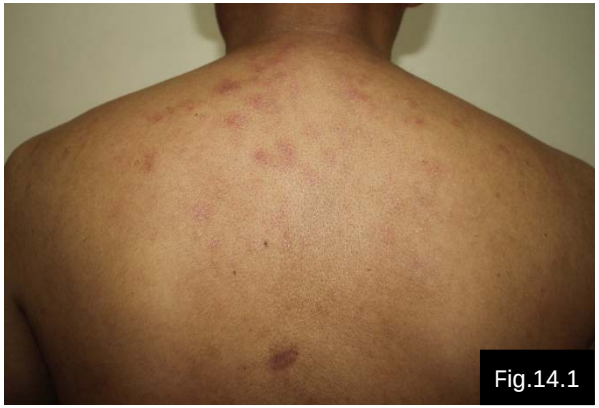
Fig.14.1

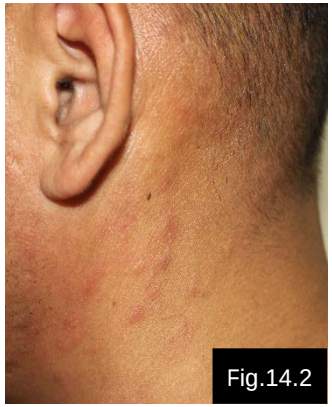
Fig.14.2

Present illness:
The patient presented with multiple itchy erythematous to brownish rashes on upper back and chest for 4 months, the rash does not relate to food, exercise, heat, or trauma. Prior to a consultation, patient was diagnosed with chronic urticaria and treated with antihistamine without improvement.

He also complained of fatigue and weight loss about 5 kilograms in 6 months. He had no fever or other systemic symptoms.

Past history: He had no underlying disease.

Family history: There was no family history of cancer or similar lesion.

Physical examination:
- No fever or lymphadenopathy
- Other systemic examinations revealed no abnormality

Dermatological examination:
- Multiple non-blanchable erythematous to brownish papules and plaques on upper back, chest, and nape of neck (Fig.14.1,14.2)

Histopathology (S19-015587, back):
- Superficial and deep perivascular dermatitis with plasma cells admixed with lymphocytes and scattered eosinophils (Fig.14.3)

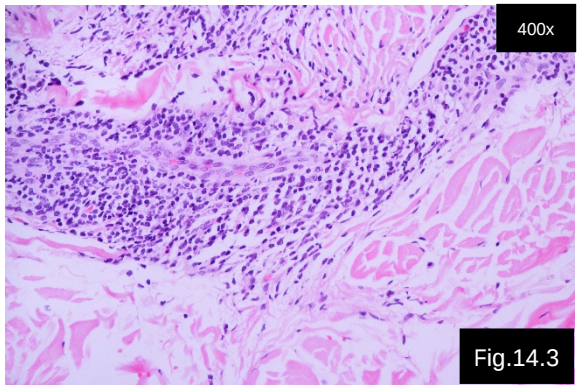

Fig.14.3

Laboratory investigations:

- CBC: Hb 14 g/dL, Hct 40.4%, Plt 433,000 /mm^3
- WBC: 10,640 /mm^3 (N 76%, L 17%, M 3%, E 3%, B 1%)
- AST/ALT: 32/40 U/L
- ALP/GGT:60/42 U/L
- TB/DB: 1/0.4 mg/dL, TP/Alb: 82.5/41.9 mg/dL
- VDRL: Non-reactive
- Serum immunoglobulin IgG: 23.410 mg/ml (N 7-16)
- SPEP: Normal pattern
- Chest x-ray: No abnormal finding

Diagnosis: Cutaneous plasmacytosis
Treatment: 0.25% desoximetasone cream apply lesions twice daily
Presenter: Taptim Stavorn, MD
Consultant: Vipawee Ounsakul, MD
Discussion:

Plasmacytosis, refers to polyclonal plasmacytic infiltrations in the skin and other organs,[1] is thought to be reactive processes that mostly occur in Asian males. It was first described by Yashiro et al[2] in 1976 and later termed *cutaneous plasmacytosis* by Kitamura et al[3] in 1980. Several years later, Watanabe et al[4] classified the *systemic plasmacytosis* as a syndrome with generalized lymphadenopathy and polyclonal hypergammaglobulinemia in

addition to the skin eruptions of cutaneous plasmacytosis. The diagnosis is based on clinicopathologic findings, the presence of a polyclonal population of plasma cells, and exclusion of secondary causes of plasmacytic infiltrations.

The skin manifestation usually consists of multiple, persistent, asymptomatic, or mildly pruritic, round-to-oval, red, violaceous to brownish macules, papules, and plaques. The lesions most commonly found on the trunk with relative sparing of the extremities. The distribution in some cases was reminiscent of a "Christmas tree" pattern.[5,6] In the large review of 69 cases by Haque et al[7], the eruption was commonly accompanied by constitutional symptoms including fatigue, malaise, weight loss, and low-grade fever. The most important of these was dyspnea and cough, which signified the lung involvement. Polyclonal hypergammaglobulinemia was observed in 88 to 93% of the cases, IgG and IgA proliferations are the most common findings.[6] Peripheral lymphadenopathy was found about 54 % of cases, usually multifocal and symmetry, involving cervical, axillary, and inguinal areas. Liver and spleen enlargement presented by up to 10 % of cases. Lung involvement found in 16% of cases, chest x-rays often showed reticulonodular or interstitial infiltration. Kidney involvement was rarely found.[7-10]

Because some cases of cutaneous plasmacytosis were found to have occult involvement of nonenlarged lymph nodes or bone marrow, the more inclusive term "*cutaneous and systemic plasmacytosis*" has been recommended.[7] In children, cutaneous plasmacytosis has been described as a distinct term called *isolated benign cutaneous plasmacytosis*, characterized by single skin lesions that consist of mature polyclonal plasmacytic cells without the systemic findings of hypergammaglobulinemia and lymphadenopathy.[11,12]

Histopathology was characterized by dense perivascular and/or periadnexal infiltration of mature plasma cells without atypia, admixed with lymphocytes and histiocytes. Many reports found that mast cell numbers were increased with some in active stages of degranulation, suggesting that they may play a role in this condition. Epidermis tended to be minimally effect, only mild acantholysis and basal hyperpigmentation were reported. Immunohistochemistry with kappa and lambda showed polyclonality of plasma cells with lack of light chain restriction.[7,13,14]

The etiology and pathogenesis of this condition have not yet been concluded; however, current evidence suggests that increased serum levels of interleukin (IL)-6 plays an important role in the pathogenesis.[8] Because of this finding, some authors proposed that cutaneous and systemic plasmacytosis is a variant of multicentric Castleman disease (MCD), a form of reactive lymphadenopathy in which increased serum IL-6 is frequently found.[15] In most cases of MCD, IL-6 appears to be produced by cells latently infected by HHV-8, whereas this virus has not been identified in any cases of cutaneous and systemic plasmacytosis by immunohistology.[16]

To date, there are only case reports or small case series for therapeutic options and no study has been performed to evaluate the different treatment modalities. Treatment options are observation, topical antibiotics, intralesional/systemic steroids, topical tacrolimus, oral doxycycline, thalidomide, antiCD-20, chemotherapy, radiotherapy, narrow band UVB, and oral/bath PUVA.[5, 14]

The majority of patients with cutaneous plasmacytosis have a chronic benign course and overall favorable prognosis. However, a few cases have demonstrated an aggressive clinical courses with a fatal outcomes.[17] Multiple extracutaneous involvements, levels of hypergammaglobulinemia greater than 5,000 mg/dL, and concentration of plasma cells in the bone marrow greater than 7% are related with a more severe disease course.[5] The development of T cell lymphoma, non-Hodgkin lymphoma, leukemia, and solid organ malignancies have been reported in patients with cutaneous plasmacytosis.[13] Whether these are coincidental findings or true associations remains uncertain. Some authors advised to do the systemic blood analysis for monoclonal gammopathy, CT/PET scan, and regular clinical monitoring at one-month to three-month intervals to look out for systemic involvement.[18]

Our patient had only fatigue and weight loss with no other systemic involvement. Blood test showed polyclonal hypergammaglobulinemia of IgG. He was diagnosed with benign cutaneous plasmacytosis and follow up is mandatory.

References:

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