Adult-onset eruptive xanthogranuloma

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ABSTRACT:

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Adult-onset xanthogranuloma (XG) is an unusual non-Langerhans cell histiocytosis which usually occurs as single lesion. Multiple lesions in adult is extremely rare which most clinical presentation affects only the skin and had no associated diseases. We reported a case of 55-year-old Thai female with multiple asymptomatic xanthogranuloma on face, neck and chest wall. Her basic laboratory investigations, chest radiography and serum protein electrophoresis (SPEP) were normal. The treatment with isotretinoin 10 mg/day was started. After 3 months of follow-up, the skin lesions were improved.

Key words: Eruptive xanthogranuloma, Isotretinoin, Histiocytosis

Case report

A 55-year-old Thai female presented with asymptomatic rashes on face, neck and chest wall for 6 months. She denied any underlying diseases. Physical examinations revealed multiple well-demarcated, dome-shaped, erythematous to orange colored papules on the face, neck and chest wall (Figure 1a-c). No mucosal involvement, lymphadenopathy or hepatosplenomegaly was identified. Other examinations were unremarkable. The ophthalmologic examination by ophthalmologist was normal. From the clinical presentations, the initial differential diagnoses were eruptive xanthoma, xanthogranuloma, and cutaneous histiocytosis and Langerhans cell histiocytosis.

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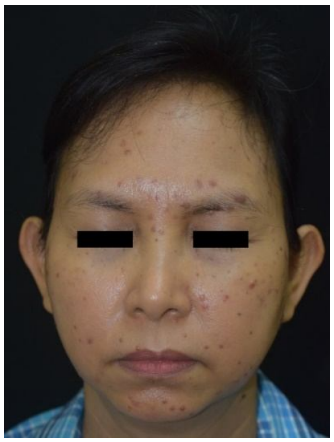

Figure 1a

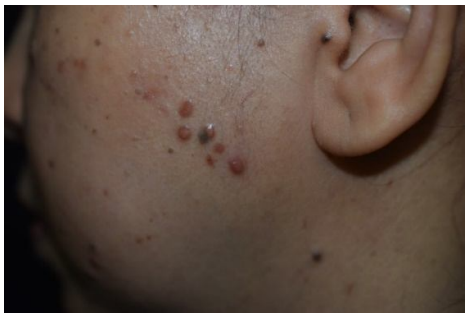

Figure 1b

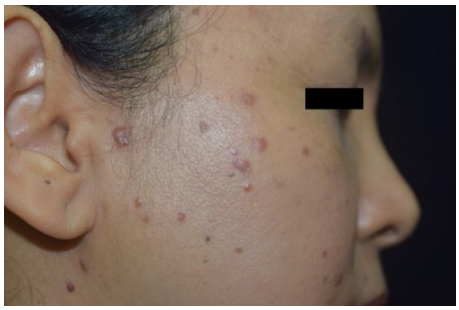

Figure 1c

Figure 1a-c Show multiple well-demarcated, dome-shaped, erythematous to orange colored papules

Skin biopsy was done from the lesion on the face. The biopsy showed nodular infiltration in upper dermis with histiocytes, foam cells, Touton-type giant cells, lymphocytes and eosinophils (Figure 2a,b). The immunohistochemical stains revealed most of the histiocytes were highlight with CD68 and Factor XIIIa and negative for S100 and CD1a (Figure 2c,d).

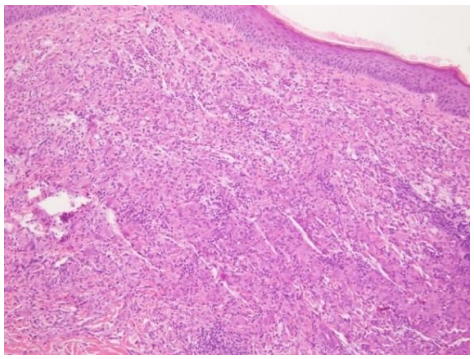

Figure 2a

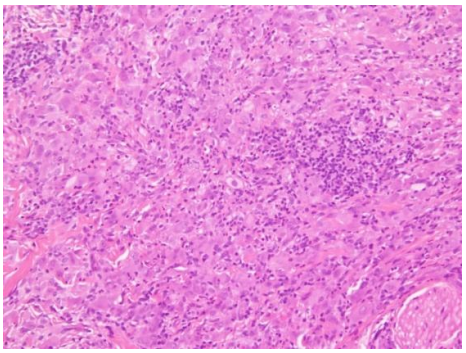

Figure 2b

Figure 2a, b Nodular infiltration in upper dermis with histiocytes, foam cells, Touton-type giant cells, lymphocytes and eosinophils (Hematoxylin and Eosin stain, X40 and X100 respectively)

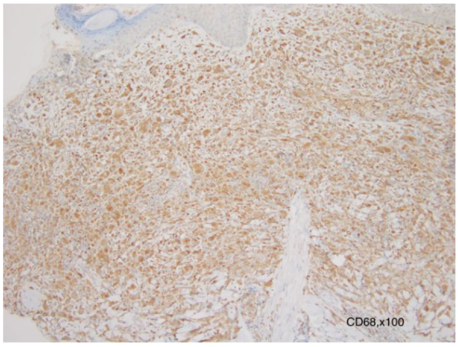

Figure 2c

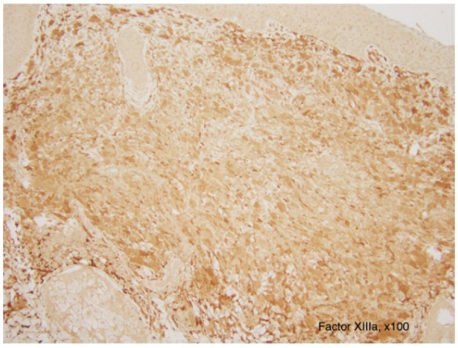

Figure 2d

Figure 2c,d Most of the histiocytes were highlight with CD68 and Factor XIIIa (CD68, X100 and Factor XIIIa, X100, respectively)

Basic laboratory investigations including complete blood count, renal and liver function test, fasting plasma glucose and lipid profile were all in normal range. Further investigation with serum protein electrophoresis (SPEP) showed polyclonal pattern. No abnormality was found on chest radiography.

The patient had final diagnosis as eruptive xanthogranuloma. The treatment with isotretinoin 10 mg/day was started. After 3 months of treatment, the skin lesions were improved (Figure 3a-c).

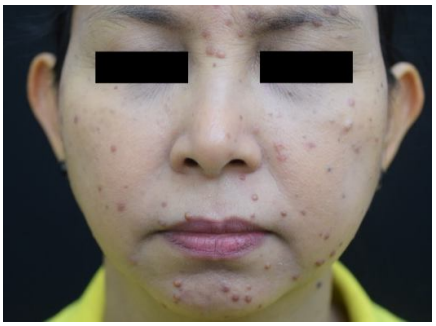

Figure 3a

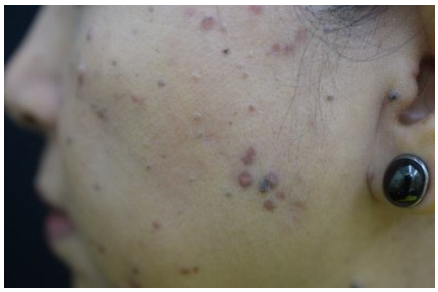

Figure 3b

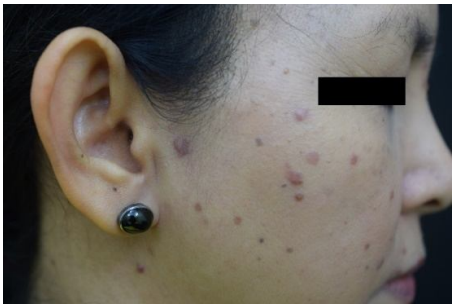

Figure 3c

Figure 3a-c After 3 months of treatment with isotretinoin 10 mg/day, the skin lesions were improved

Discussion

Xanthogranuloma (XG) is a benign non-Langerhans cell histiocytosis which around 80% of cases, lesions appear in the first year of life. Adult onset XG is rare and usually occur as single lesion in late twenties and early thirties[1,2].

Multiple lesions in adult is extremely rare which defined as presence of more than 5 XG lesions in patient older than 14 year of age[2-4]. This type is common reported in males with average age of diagnosis is 46.7 years[3,4].

The etiology is unknown, possibly reactive histiocytes response to stimuli including trauma, infections and neoplasms[1]. Additionally, the uptake of low-density lipoprotein and the synthesis of cholesterol within macrophages are increase in adult with XG which may explain the accumulated cholesterol intracellularly in the absence of hyperlipidemia[5].

In general, cutaneous XG has no genetic mutation[6]. Nevertheless, few cases of extracutaneous pediatric intracranial XG with BRAF V600E mutation have been reported[7].

There was a single case report of BRAF V600E mutation in adult cutaneous XG coexistence with Langerhans cell histiocytosis. However, the mutation was identified only in the Langerhans cell populations and it was not found in the histiocytes population in the tumor[8]. Beside BRAF mutation, mitogen-activated protein kinase -1 (MAPK-1) gene mutation was also identified in a 10-year-old boy with atypical aggressive juvenile XG. This patient presented with both cutaneous and multiple internal organ involvements[9]. Until now, there is no report of genetic mutation in adult cutaneous XG[10,11].

The clinical manifestations in adult are similar to childhood. The lesions presented with yellow to brown-colored domed-shaped papules and nodules predilection on the upper body, especially head and neck regions[12-14].

Generally, extracutaneous involvements in children with juvenile XG have been mentioned which including eyes, soft tissue and internal organs such as lungs, liver, spleen, testes, pericardium, gastrointestinal tract, kidney, and central nervous system[15]. Additionally, children with juvenile XG and neurofibromatosis type I have higher risk for develop chronic myeloid leukemia[1,12].

In contrast to juvenile XG, very few case reports of adult XG had extracutaneous involvements. In these reports, each patient had XG on the breast and iris without any cutaneous lesions[16-17].

Most of the adult multiple XG had no underlying diseases[12-15]. Nevertheless, a number of associated diseases had been identified including follicular lymphoma, chronic lymphocytic leukemia, large B-cell lymphoma, essential thrombocytosis, monoclonal gammopathy, non-insulin-dependent diabetes

mellitus, phyllode breast tumor and gastrointestinal stromal tumor[4,12,13,18-20].

The histopathologic features of XG show well-demarcated nodular infiltration in the dermis with histiocytes. Foamy appearance histiocytes with Touton giant cells are the typical finding and usually present in mature lesion. Other inflammatory cells including lymphocytes, eosinophils and plasma cells are also found in the lesion[1]. The immunohistochemical stains with histiocyte markers such as CD68, HAM-56 and factor XIIIa highlight the tumor cells. S100 protein can be expressed in some case whereas CD1a and Langerin (CD207) are negative[1,14,21].

Unlike juvenile XG, spontaneous regression is rarely reported in adult XG. Only single case of adult multiple XG had spontaneous resolution after 1 year of follow up period[22].

Multiple treatment modalities were mentioned including cryotherapy, surgical excision and carbon dioxide laser[13,21]. Medical treatments with 2 months of oral isotretinoin 20 mg/day can also improve the cutaneous lesions[13,15]. In patients with associated hematologic diseases, treated underlying diseases also improved the cutaneous XG lesions[18-20].

In conclusion, we report a case of adult onset eruptive xanthogranuloma without underlying disease or systemic involvement. The lesions improved with oral isotretinoin 10 mg/day for 3 months.

Although associated diseases or systemic involvement is fewer reported in adult XG compared to juvenile XG, periodically follow up is still required.

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