

A case of adult onset disseminated juvenile xanthogranuloma

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ABSTRACT

Juvenile xanthogranuloma (JX) is a rare, benign, non-Langerhans histiocytic proliferative disease that etiology is unknown. It is usually seen in children and infants. JX in adult is very rare. A 41-year-old female patient was admitted to our clinic with papules on her face, torso and extremities. A few lesions had occurred 3 years ago on her face, they disseminated all over her body after having a traffic accident one year ago which for she had operations and she also concurrently was diagnosed as diabetes mellitus (DM). Based on clinical and histopathological findings, the diagnosis of JX was made. There is no systemic involvement of JX detected. JX seen in adults are very rare and usually associated with hematological malignancy. The present case is a rare adult onset disseminated JX case without malignancy association and representative for the opinion that trauma and DM may be triggering factors.

Key words: Non-Langerhans histiocytosis; Juvenile xanthogranuloma; Adult

INTRODUCTION

Juvenile xanthogranuloma (JX) is a rare, non-Langerhans histiocytic proliferative disease, generally seen in childhood [1]. JX in adult is very rare and in some of adult cases, there were reported concurrent hematological malignancies [2-4]. In the present case, a 41-year-old female patient who has disseminated JX lesions with adult onset is presented.

CASE REPORT

41 year old female patient was admitted to our clinic with papules all over her body. Lesions started 3 years ago on her face and they remained stable until having a traffic accident 1 year ago. After accident and subsequently operation, the lesions began spreading on whole body. In her personal history, there were a surgical

operation because of the accident and concurrently diagnosis of diabetes mellitus. In her family history there was nothing important. In dermatological examination, multiple yellowish-pink coloured, some dome-shaped, indurated papules were detected on her head, hairy skin, external auditory canal, pubis and upper torso (Figs 1 - 3). Routine laboratory tests were normal. Histopathological examination of the lesions showed histiocytic infiltration and multinuclear giant cells and actin, desmin, S100 and factor XIIIa stains were found negative whereas CD68 was positive (Figs 4a - d). Based on clinical and histopathological findings, patient was diagnosed as JX. There were no pathological findings in ocular and respiratory examinations of the patient. The patient was followed because the lesions has possibility of spontaneous resolution. But, 6 months later, there were no regression in control examination.

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Figure 1: Multiple yellowish-pink coloured, some dome-shaped, indurated papules on the torso.



Figure 2: Multiple yellowish-pink coloured papules on the scalp.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

JX is an uncommon non-langerhans histiocytic proliferative disease which 75% of cases were diagnosed in the first year of life. It is rarely seen in adults and the lesions generally seen as solitary and resisted to spontaneous regression [1]. Some of the adult cases with disseminated lesions were associated hematologic malignancy, mostly leukemias [2-4]. Our case is adult onset disseminated JX case without malignancy association.

The etiology of JX is still unknown however, it is suggested that it might be a reactive disorder responding to any traumatic or infectious stimulus [1].



Figure 3: Papule on external auditory canal.

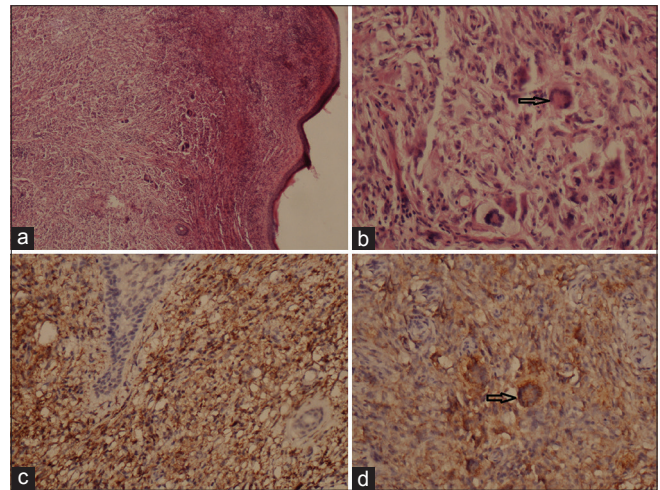


Figure 4: a) Histiocytic infiltration and multinuclear giant cells (H&E x40), b) Closer apperance arrow: Touton type giant cell (H&E x200), c) CD68 positivity of histiocytes (CD68x 40), d) Closer apperance arrow: Touton type giant cell (CD68x200).

Our case has trauma history and diabetes mellitus and dissemination of the lesions was simultaneously with trauma diagnosis of diabetes mellitus. So, we think that trauma and/or diabetes mellitus may be potential triggering factors for JX.

In many cases, lesions are limited to the skin and benign characterized [1]. Extracutaneous infiltration was reported in 5% of cases. The most common site are eye and lung. Early recognition is important to avoid complications [5]. In our case, there was no pathological findings in physical or radiological examinations that indicates any systemic involvement.

Associations between JX and neurofibromatosis type 1 (NF1), juvenile myelomonocytic leukemia (JML), urticaria pigmentosa, Niemann-Pick disease and

diabetes mellitus were reported [6]. Our case has also diabetes mellitus.

Differential diagnosis firstly includes non Langerhans-Langerhans cell histiocytosis. The other differential diagnoses are xanthomas, molluscum contagiosum, neurofibroma, spitz nevus. Diagnosis of JX is done by clinical, histopathological and immunohistochemical examinations [7,8]. In histological examination of JX lesions, characterized Touton cells with ring shaped nucleus, foamy cytoplasm are seen and S100, CD1a stains are negative whereas CD68 stain is positive similarly in our case [9].

The lesions usually regress spontaneously in 3-6 years or sometimes with hyperpigmentation, atrophy or anetoderma. For diagnosis or cosmetic concerns, they could be excised [1]. Our case has multiple lesions, so excision was not suggested.

In conclusion, JX may occur in adult and dermatologist should keep in mind in the patients with yellowish-pink coloured, indurated papules. Trauma and diabetes mellitus may be a triggering factor for dissemination of JX.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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