

Diffuse juvenile xanthogranuloma: A case report

Hafsa EL Boukili*; Meryem Soughi; Zakia Douhi; Sara Elloudi; Hanane Baybay and Fatima Zahra Mernissi

Department of dermatology University Hospital Hassan II, Fez, Morocco.

Faculty of Medicine and Pharmacy, Sidi Mohamed Ben Abdellah University, Fez, Morocco

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***Corresponding Author:** H EL Boukili, Department of dermatology University Hospital Hassan II, Fez, Morocco.
E-mail: hafsaboukili@gmail.com & hafsa.elboukilielmakhoukhi@usmba.ac.ma

Abstract

Diffuse juvenile xanthogranuloma (DJX) is a rare non-Langerhans histiocytosis characterized by the presence of multiple yellowish papules and nodules on the skin. It mainly affects children and young adults, with a slight predominance in males [1]. The lesions can occur anywhere on the body but are commonly found on the face, neck, and trunk. They are usually painless and may be asymptomatic. DJX can also involve other organs, such as the liver, spleen, and bone marrow, although this is less common. The exact cause of DJX is not well understood, but it is thought to be related to an abnormal immune response [2]. We report here a new case of diffuse juvenile xanthogranuloma.

Keywords: Xanthogranuloma; Juvenile; Diffuse.

Introduction

Juvenile xanthogranuloma (JXG) is a benign non-Langerhansian histiocytosis that classically presents as yellowish skin nodules due to accumulation of lipid-laden macrophages. It is usually unique but multiple or even disseminated forms have been described. We report here a new case of diffuse juvenile xanthogranuloma.

Observation

A 17-month-old female, with no previous pathological history, She was born at 40 weeks' gestational age, without any abnormality or history of trauma at birth. Presented with a diffuse asymptomatic eruption of progressive onset since the age of 4 months. Dermatologic examination revealed well-limited, firm, infiltrated, rounded to oval, orange papules that ranged from 0.5 to 2 cm in diameter on the face, trunk, upper and



Figures 1-2-3: Clinical features of skin lesion. Diffuse crimson papules on the face, trunk and extremities.

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lower extremities (Figures 1, 2 and 3) sparing the palmoplantar region and without mucosal involvement. Darier's sign was negative, there was no milky coffee spot or other associated skin lesion and no palpable adenopathy. Immunohistochemistry yielded results immunoreactive for CD68, weak focal positivity for S-100 protein, but showed negative for Langerin. These results confirmed that the histiocytes are non-Langerhans cells. There was no other systemic involvement or evidence that could point to NF1. Spontaneous regression was observed, justifying therapeutic abstention with close clinical monitoring.

Discussion

Juvenile xanthogranuloma (JXG) is a rare proliferative skin disorder in the group of non-Langerhans cell histiocytosis [3], it typically occurs in infancy or early childhood, although adult-onset JXGs have been reported [4]. JXG may be present at birth, but it mostly arises during the first year of life [5]. It is characterized by one or more nodules with the predilection sites on the head and neck, although the appearance on the trunk, extremities and extracutaneous locations has been reported also [6]. Skin lesions are self-limited and vary in size. They are reddish or yellowish benign papules or nodules, histologically, JXG is composed of collections of histiocytes, foamy cells and Touton giant cells. The diagnosis of JXG is mainly clinical, but sometimes a biopsy analysis is required [7]. While JXGs classically present as a solitary entity, they can also manifest as multiple lesions. Single JXGs typically present in the first year of life, while multiple JXGs most commonly occur as congenital lesions or in the first six months of life. Whereas a single JXG can often be diagnosed by clinical presentation, multiple JXGs are more challenging because they can mimic disorders such as Langerhans cell histiocytosis (LCH), molluscum contagiosum, and pyogenic skin infection. The confirmation of clinical diagnosis can be made by skin biopsy [8].

In these cases of multiple XGJ, rare extracutaneous involvement has been reported. The most common is ocular involvement, particularly hyphaema, in children under 2 years of age. Other extracutaneous locations of JXG are the lung, heart, gastrointestinal system, central nervous system, adrenal gland, pituitary gland, bones, bone marrow, and kidney [6].

Apart from complications related to extracutaneous involvement, the prognosis is generally good and most cases followed a benign course with spontaneous resolution, within a few months or years, therefore a progression to fatal complications is possible. The cases resulting in fatality had wide spread involvement of multiple organ systems and developed sequelae including liver failure and sepsis, coagulopathy, and electrolyte disturbances liver failure and sepsis, coagulopathy, and electrolyte disturbances [8].

Conclusion

JXG is a self-limited condition. Skin lesions usually resolve spontaneously. Treatment is necessary in those with extracu-

taneous involvement who may have an increased morbidity. JXG is rarely multiple, which is the originality of our observation, illustrating a clinically atypical observation of JXG by the multilesional and multifocal nature of lesions.

References

1. Takahashi S, Muto J, Takama H, Yanagishita T, Takeo T, Ohshima Y, et al. Generalized eruptive histiocytoma: Pediatric case report and review of the published work. *J Dermatol.* 2019; 46: 407-8.
2. Placide N, Nicolas S. Mitotically active xanthogranuloma: a case report with review of the literature. *Am J Dermatopathol.* 2012; 34: 27-30.
3. Dehner LP. Juvenile xanthogranulomas in the first two decades of life: a clinicopathologic study of 174 cases with cutaneous and extracutaneous manifestations. *Am J Surg Pathol.* 2003; 27(5): 579-593.
4. Hernandez-Martin A, Baselga E, Drolet BA, Esterly NB. Juvenile xanthogranuloma. *J Am Acad Dermatol.* 1997; 36(3-1): 355-359.
5. Jung T, Emmert S, Günzl HJ, Neumann C, Rüniger TM: Congenital manifestations of juvenile xanthogranuloma (large nodular form). *Hautarzt.* 2000; 51: 423-426.
6. Pajaziti L, Hapçiu S, & Pajaziti A. Juvenile xanthogranuloma: a case report and review of the literature. *BMC Research Notes.* 2014; 7(1): 174. doi:10.1186/1756-0500-7-174.
7. Sangüeza OP, Salmon JK, White CR Jr, Beckstead JH: Juvenile xanthogranuloma: a clinical, histopathologic and immunohistochemical study. *J Cutan Pathol.* 1995; 22(4): 327-335.
8. So N, Liu R, & Hogeling M. Juvenile xanthogranulomas: Examining single, multiple, and extracutaneous presentations. *Pediatric Dermatology.* 2020.