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Giant and Profuse Xanthogranuloma in a 14-Month-Old Infant at the Dermatology Hospital of Bamako

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Abstract **Case Report**

Juvenile xanthogranuloma (JXG) is a proliferative disease included in the group of non-Langerhansian histiocytoses (NLH) that mainly affects infants and children, and is congenital in 40-70% of cases. This condition is usually a single or few lesions but the profuse form is very rare. We report a case of giant, profuse xanthogranuloma in a 14-month-old infant. A 5-month-old infant was brought to the dermatology department with a rash that had been flaring for more than a month. The infant had no previous history of the disease and the lesions were asymptomatic. Clinical examination revealed firm nodulopapular lesions of normal skin colour, varying in size from 5 mm to 1 cm, scattered over the body. The lesions were most prominent on the neck and trunk. Biopsy of one nodule was concluded to be a juvenile xanthogranuloma. We reassured the parents about the disease and abstained from treatment. After 9 months of evolution, the child was readmitted to dermatology for aggravation of the rash. On examination, turgid nodules of 1-2cm in diameter were observed, profuse on the body. Elsewhere the clinical and biological examination was unremarkable. We report a case of giant diffuse xanthogranuloma in a 14-month-old infant, evident by histological examination. Our observation is peculiar because of its mode of extension and the giant nature of the lesions. It remains to be seen whether the evolution of this disease passes through this turgid stage. Further work is needed to support the pathogenesis of the lesions.

Keywords: Xanthogranuloma, giant, infant, Dermatology Hospital, Bamako.

INTRODUCTION

Juvenile xanthogranuloma (JXG) is a proliferative disease included in the group of non-Langerhansian histiocytoses (NLH) that mainly affects infants and children; this condition is congenital in 40-70% [1]. The dermatological presentation is classically papular or single or multiple nodular lesions of widely varying size ranging from a few millimetres to several centimetres, firm with a smooth surface. Initially flesh-coloured and then reddish- brown, they gradually take on a typical orange-yellow colour [1]. Atypical, lichenoid, reticulated, maculo-papular, plaque-like or linear skin forms have been described from isolated observations. JXG with multiple lesions, characterised by more than five lesions, is a rare presentation with lesions usually regressing within 1-5 years [2, 3]. The condition is benign and usually heals in adulthood without sequelae.

We report a case of a giant, profuse xanthogranuloma in a 14-month-old infant.

OBSERVATION

A 5-month-old male infant from a full-term pregnancy, with no other known pathological history, EPI vaccine up to date.

The child was brought by his mother to the dermatology department for a rash that had been evolving in flare-ups for more than a month. Clinical examination revealed firm nodulopapules of normal skin colour, varying in size from 5 mm to 1 cm, scattered over the body. The lesions were most prominent on the neck and around the seat, asymptomatic. Biopsy of one nodule was concluded to be a juvenile xanthogranuloma (a very dense histiocytic infiltrate in the dermis and so-called Touton giant cells).

We reassured the parents about the disease and made a therapeutic abstention.

After 9 months of evolution the child was readmitted to dermatology for worsening of the rash. On examination, turgid nodules of 1-2cm in diameter were observed, profuse on the body. Elsewhere, the clinical examination showed lymph nodes in particular. The biological work-up was unremarkable: normal tryptase, fasting blood sugar and lipid levels. The ophthalmological examination was normal.

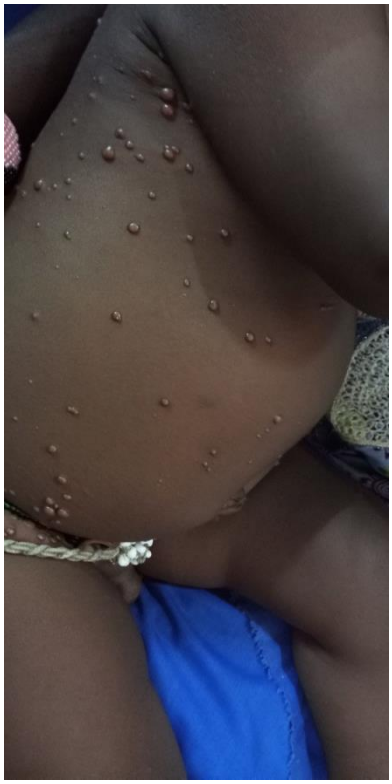

Image 1: 5-month-old infant (multiple diffuse papulo-nodual lesions)

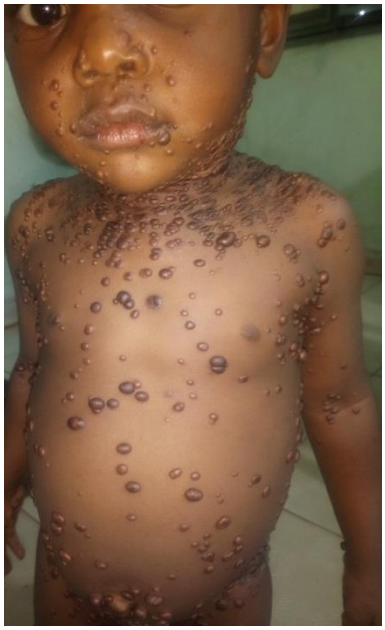

Image 2: A 14-month-old infant (multiple diffuse brownish nodular lesions)

DISCUSSION

We report a case of a giant diffuse juvenile xanthogranuloma in an infant, evident by the clinical aspects, asymptomatic skin-coloured papulo-nodules but also on histology by the presence of histiocyte-rich infiltrate and so- called Touton giant cells.

XGJ is a non-Langheransian histiocytosis, a rare disease of unknown aetiology, usually benign, first reported by Adamson in 1905 as congenital Xanthoma multiple, corrected in 1954 by description of cellular features under the microscope.

A common condition in children, present at birth in 5-17%, about 70% of XGJs occur in children under one year of age, making it a childhood condition [6], although cases have been observed in adults as well as in men and women [5].

The main clinical manifestation is a skin-coloured, yellow-orange, papulo-nodular lesion from a few millimetres to several centimetres in diameter. The lesion is mostly cutaneous and single but sometimes multiple, occurring preferentially on the neck, trunk and extremities. Extracutaneous manifestations are uncommon, mainly dominated by ocular involvement [4]. This information is consistent with the observations made in our patient; a 5-month-old infant at the onset of the eruption, multiple lesions scattered over the body, asymptomatic skin colour, but peculiar in its mode of extension, the giant and turgid nature of the lesions.

Some authors report that multiple cutaneous forms of XGJ in children under 2 years of age were associated with a high risk of extracutaneous lesions, particularly ocular, pulmonary, cardiac and gastrointestinal [4, 6].

The diagnosis of XGJ can be clinical, given the low incidence of the disease, the clinical resemblance with certain diseases such as mastocytosis, noevus spitz, the contribution of the physician to the clinical diagnosis proves difficult according to certain authors, which increases the diagnostic errors sometimes corrected by histology [5]. The histological characteristics depend on the stage of the lesion. It is very easy to distinguish in the granulomatous stage where a very dense histiocytic infiltrate in the dermis and giant cells called Touton cells can be observed. Touton cells are multi-nucleated giant cells with a homogeneous eosinophilic appearance in the centre of the cytoplasm and peripheral xanthomisation as in our case. In the early stage of the disease, this xanthomisation or Touton's cells may be missing, which sometimes makes it difficult to distinguish from Langheran histiocytosis and justifies the use of immunohistochemistry [4]. Histiocytes in XGJ are predominantly immunoreactive with KiM1P or CD68, factor XIIIa and non-immunoreactive with CD1a and protein S100. In contrast, cells in langheran histiocytosis are immunoreactive with S100 protein, CD1a and non-immunoreactive with CD68 and factor VIIIa.

CONCLUSION

Xanthogranuloma is genomically a unique lesion. Our observation is particular by its mode of extension and the giant character of the lesions. It remains to be seen whether the evolution of this disease passes through this turgid stage. Further work is needed to support the pathogenesis of the lesions.

REFERENCES

1. Samuelov, L., Kinori, M., Chamlin, S. L., Wagner, A., Kenner-Bell, B. M., Paller, A. S., ... & Mancini, A. J. (2018). Risk of intraocular and other extracutaneous involvement in patients with cutaneous juvenile xanthogranuloma. *Pediatric dermatology*, *35*(3), 329-335.
2. Ferreira, B. R., Cardoso, J. C., Reis, J. P., & Tellechea, Ó. (2017). Multiple adult-onset xanthogranuloma, an uncommon diagnosis. *Anais Brasileiros de Dermatologia*, *92*, 294-295. doi:10.1590/abd1806-4841.20176216
3. Pradhan, S., Chen, S., & Xiong, L. (2017). Rapports de cas sur le xanthogranulome juvénile et brève revue de la littérature. *Science*, *6*(5), 82–85.
4. Pajaziti, L., Hapçiu, S. R., & Pajaziti, A. (2014). Juvenile xanthogranuloma: a case report and review of the literature. *BMC research notes*, *7*, 1-3.
5. Dirk, J., & Dieter, H. (2005). Juvenile xanthograluma in childhood and adolescence. In *Am. J Surg. Pathol.*, *29*, 21-28.
6. Cypel, T. K. S., & Suker, R. M. (2008). Juvenile xanthogranuloma : a case report and review of literature. In *Can J Plast Surg.*, *16*(3), 175-177.