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Juvenile Xanthogranuloma: A Case Report with Literature Review

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Abstract

Juvenile xanthogranuloma (XGJ) is the most common form of non-Langerhansian histiocytosis. We report a pediatric case of multiple XGJ without visceral involvement in a 6-month-old female infant who was hospitalized with disseminated congenital skin nodules, of firm consistency, of variable diameter ranging from a few millimetres to 1 cm, involving the face, scalp, trunk and limbs. There was no mucosal localization. The remainder of the somatic examination was unremarkable. The diagnosis of disseminated XGJ was confirmed by skin biopsy without visceral or systemic involvement. Therapeutic abstention was recommended. This observation underlines the fact that XGJ is a rare clinical entity that should not be overlooked when faced with congenital skin nodules.

Keywords

Cutaneous Nodules, Juvenile Xanthogranuloma, Child

1. Introduction

Juvenile xanthogranuloma (XGJ) is the most common form of non-Langerhansian histiocytosis [1]. It is a benign, self-invasive tumor. Forming a single lesion in more than two-thirds of cases, XGJ can also have disseminated lesions. We report a pediatric case of multiple XGJ without visceral involvement.

2. Observation

A 6-month old female infant of non-inbred parents, the youngest of four siblings, was admitted for the management of skin nodules they were present from birth and changed gradually into larger, evolving in a context of apyrexia and conservation of the general state. In the antecedents, a well-monitored pregnan-

cy was noted, estimated at term, birth weight of 4 kg, good psychomotor development. Clinical examination showed brownish nodular lesions on healthy skin, of firm consistency, of variable diameter ranging from a few millimetres to 1 cm, involving the face (**Figure 1**), scalp (**Figure 2**), trunk (**Figure 3**) and limbs. There was no mucosal localization. The ganglion areas were free, there was no hepato splenomegaly. Paraclinical explorations (blood count, liver tests, renal function) showed no abnormalities. The ophthalmologic examination was unremarkable.

The diagnosis of XGJ, strongly suggested clinically, was confirmed by skin biopsy. Anatomopathological examination showing a dense, well defined dermal cell infiltrate composed of histiocytes of variable size and shape, and an inflammatory granuloma composed of lymphocytes, eosinophilic and sometimes neutrophilic polynuclear cells, multinucleated Touton-type giant cells. A further immuno-histochemical study was carried out showing tumour cells expressing CD163, CD68, factor XIIIa. They were negative for CD1a and PS100. The evolution was spontaneously favorable.

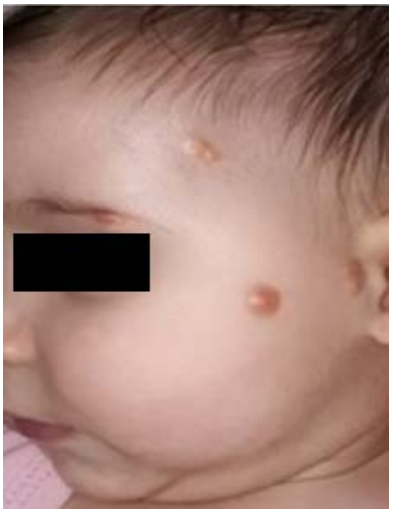

Figure 1. Facial skin nodules.

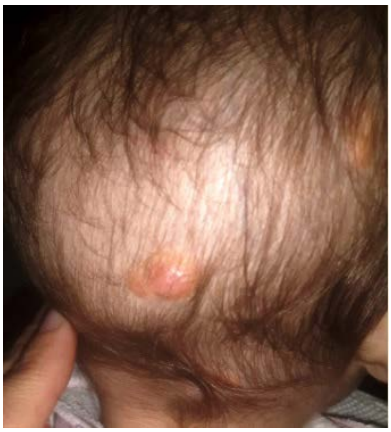

Figure 2. Skin nodules on the scalp.

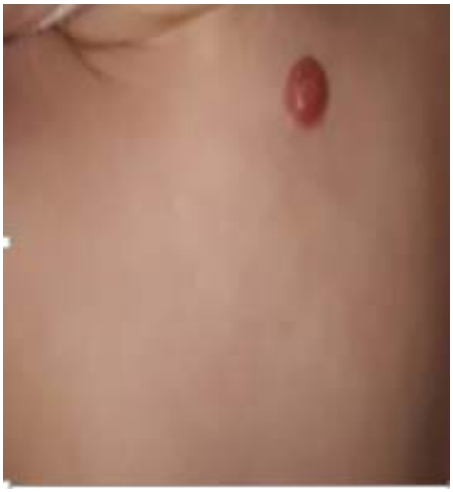

Figure 3. Skin nodules on the trunk.

3. Discussion

XGJ is a relatively common form of non-Langerhansian histiocytosis in small children. XGJ is present from birth in 5% to 17% of cases as in our case and appears in 40% to 70% of cases during the first years of life, but it can also occur in adulthood [1]. Clinically, XGJ is a well-defined, firm, round or oval, yellow-orange, brown, pink or red papule or nodule. Its size varies from 0.5 to 2 cm in diameter. Some telangiectasia may be present on the surface.

Semiologically atypical forms include lichenoid, reticulated, maculopapular, plate-like, linear eruptions. It is usually asymptomatic. In 60% of cases, the lesion is single, but multiple or even disseminated forms have been reported. Multiple disseminated lesions may be associated in 4% of cases with visceral localizations, sometimes severe [2]. Preferred areas of involvement are the head and neck, trunk and limbs but all locations were reported including mucous membranes [1]. There are two forms of XGJ: the small nodule form and the large nodule form. Our patient had a mixed form with lesions of different sizes and shapes. The mucous membranes may be affected and in some cases there is systemic dissemination [2] [3]. Half of the systemic forms of XGJ are associated with skin involvement. The size and number of skin lesions do not correlate with systemic manifestations [4]. In patients with multiple XGJDs, visceral involvement should be sought if there is clinical evidence of referral. In this case, brain CT or MRI, chest X-ray, abdominal CT or ultrasound, bone scan or bone imaging, ophthalmologic examination and possibly bone marrow biopsy should be performed [2]. Extracutaneous damage is very rare, with ocular damage being the most common extracutaneous location. Its incidence is 0.3% - 0.5% in patients with skin lesions and 41% of patients with eye involvement have multiple skin lesions. The risk of ocular injury is greatest in the first two years of life [3] [4] and in children with multiple skin lesions. During ocular injury, the iris is the most common location. It is manifested by localized or diffuse iris tumor, unilateral glaucoma, spontaneous hyphema, signs of uveitis, and congenital or

acquired heterochromia. The eyelid is the second location after the iris [5]. Other extracutaneous injuries are very rare: pulmonary, hepatic, and even more rarely splenic, pancreatic, retroperitoneal, digestive, renal, gonadal, cardiac, and nervous [1].

XGJ are more frequently reported with café au lait spots, whether or not associated with epilepsy, and a family history of neurofibromatosis type 1 (NF1) [6]. The association of multiple XGJ with NF1 affects 18% of patients [1]. It must be known, as acute myelomonocytic or chronic monocyticleukaemia may occur in these forms [3]. Zvulunov *et al*. estimated that patients with NF1 and multiple XGJ would be 20 - 30 times more likely to develop chronic juvenile myelomonocyticleukaemia than those with isolated NF1 [1] [7]. Our patient did not have café au lait spots, a family history of NF1 or abnormal blood counts. The characteristic histopathological image is that of a dense, well defined dermal cell infiltrate composed of histiocytes of variable size and shape, and an inflammatory granuloma composed of lymphocytes, eosinophilic and sometimes neutrophilic polynuclear cells, multinucleated Touton-type giant cells. Immunohistochemistry makes it possible to distinguish XGJ from Langerhans cell histiocytosis in particular; XGJ histiocytes express CD163, CD68, factor XIIIa but neither PS100 nor CD1a. Histiocytic proliferations of nodular presentation should be considered in differential diagnoses. Hashimoto-Pritzker histiocytosis is often ulcerated and usually regresses rapidly within a few months. In immunohistochemistry, PS100 labelling is positive for 10% to 20% of cells. Benign cephalic histiocytosis is sometimes impossible to distinguish histologically from an early form of XGJ. However, lesions are flatter in benign cephalic histiocytosis and reach the head and neck. Clinically, Spitz nevus can be difficult to distinguish. Histological examination will show a proliferation of spindle epithelioid cells.

For prognostic and therapeutic reasons, non-Langerhans cell histiocytosis, which are usually benign and self-involving, must be distinguished from the forms langerhans, potentially serious, characterized by specific expression of the CD1a and CD207/langerine antigens. In electron microscopy, histiocytes of a Langerhans nature contain characteristic cytoplasmic inclusions: Birbeck granules.

The prognosis is generally benign with a regression in children in three to six years according to the data in the literature; a priori there is no regression in adults [1].

After involution, sequelae such as hyperpigmentation and atrophy may be observed. Recurrences after resection have also been reported.

4. Conclusion

This observation underlines the fact that XGJ is a rare clinical entity not unknown in front of congenital skin nodules and that clinical examination should focus on the search for extracutaneous damage and signs of NF1 with a systematic ophthalmologic examination are justified in case of extracutaneous dam-

age or multiple lesions.

Conflicts of Interest

The authors declare no conflicts of interest.

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