Cutaneous Horn, Juvenile Xantho Granuloma in the First Decades of Life, A Clinico-Dermatoscopic Correlations

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Abstract

Background: Juvenile xanthogranuloma (JXG) is a rare disorder that present at birth, in infancy or in adults. It can be either solitary or multiple nodules of various sizes. It can be associated with internal organ involvements especially eyes, lung and liver consecutively. The diagnosis can be made with confidence by clinical examination alone and confirmed by histology in doubtful cases.

Hereby I report the useful benefit of dermoscopy in evaluating and diagnosing a skin lesion that suggestive of JXG.

Case presentation: One year white girls presented with gradual onset of a nodular lesion on her forearm that shown a surface projecting horn. A biopsy was planned and scheduled. Clinical features were in keeping with JXG.

Conclusion: Juvenile xanthogranuloma (JXG) is on the spectrum of histiocytosis variants which should be examined thoroughly in children to exclude underlying malignancy. The purpose of this article is to describe the clinical dermatoscopic correlated features of the JXG. This is interesting case of JXG presenting in a young white Swedish female child.

Keywords: Juvenile xanthogranuloma (JXG); Nodule; Dermatoscopy; Dermoscopy; Child

Introduction

Juvenile xanthogranuloma (JXG) is a rare benign, non-Langerhans histiocytosis, macrophages loaded with lipids, self-healing, and skin lesion that is predominantly affecting infants and children. It can be either solitary or multiple yellowish or red-brown, asymptomatic firm papules or nodules affecting trunk or limbs without a preceding trauma and can resolve spontaneously within several years with black male predominance however this stays to be arguable [1]. The main differentials can be Molluscum contagiosum or viral warts, mastocytomas, spitz nevi, leishmaniasis, tuberculosis, and Langerhans’ cell histiocytosis, reticulo-histiocytoma and sometimes pyogenic granuloma.

Histology usually reveals hyperkeratosis and parakeratosis in the epidermis and collection of histocytes with infiltration in the dermis with foamy and multinucleate Touton giant cells. The diagnosis is mainly clinical, however sometimes a biopsy reading is necessary.

JXG is the variant of non-Langerhans’ cell histiocytosis. The orange-yellow background coloration with cloud of pale yellow globules is the most distinctive dermoscopic finding of fully developed and regressed JXG stages, with erythematous margin, faint pigment network and white linear streak.

Case Presentation

A healthy one-year-old Swedish female child with onset in eighth month of life presented with a pimple like skin lesion on the right forearm mid flexor at the clinic of dermatology (Figure 1). She was born without any morbidity. The parents reported that in the last four months it was growing steady and developed a surface crust without ulceration that enlarged and expanded gradually into an upward projecting horn (2 × 1). Past medical history is unremarkable (Figure 2). Examination revealed a firm conical shaped lesion with brownish orange-red hue, 5 mm in diameter and 3 mm high. The child was otherwise healthy, feeding well. And no other skin lesion can be found.
Figure 1 Right mid flexor forearm showing hard conical shaped lesion with brownish red hue.

Figure 2 Upward projecting cutaneous horn.

Figure 3 Dermatoscopy showing a well circumscribed skin lesion with yellowish white clots and linear and branched vessels were predominantly at the periphery of the lesion at one pole on an orange-yellow background.

Dermatoscopy conducted by Heine Delta 20 non-polarized contact, and it revealed a well circumscribed skin lesion with yellowish white clouds and linear and branched vessels were predominantly at the periphery of the lesion at one pole on an orange-yellow background (Figure 3).

A surgical planned excision biopsy was given however; total spontaneous involution had happened insidiously and thus histology diagnosis was not verified.

Discussion

JXG is the most common non-Langerhans cell histiocytosis of childhood [1,2].

These lesions are normally asymptomatic and disappear spontaneously within 3 to 6 years. They are a type of granulomatous process and the actual cause is unknown, however some postulation of an underlying injury or infection that causes macrophage responded disorderly. It has been stated that children younger than two and with multiple nodules are at risk for ocular involvement. Any organ can be involved with JXG, however the commonest in order are eyes, lungs, and liver consecutively.

JXG was first coined in 1905 by Adamson. Dermoscopic appearances of JXG were first described by Palmer and Bowling. Since then, few cases have emphasized the clinical usefulness of dermoscopy.

Cutaneous horn is characteristic in such cases which can be similar to an animal horn. Horns can be seen in other similar conditions where it could need a through workup to distinct between them. Horns can be seen in premalignant cases such as actinic keratosis (AK) and Bowen disease (BD), Molluscum contagiosum (MC), Xanthomatous dermatofibroma (XDF), and in Seborrhieic keratosis (SK) and filiform warts and malignant cases such as Keratoacanthoma base (KA) and squamous cell carcinoma (SCC). Thus, finding out the underlying disease is paramount to send for histology sign out.

JXG is not a true pathological condition, however a reactive one with keratinization.

JXG has three distinctive stages [1] Early evolutionary stage [2] Fully developed stage and [3] Late regressive stage [4] Dermoscopic features constitute some main features. The setting sun appearance which can be seen mostly in the first
two stages of the JXG, then in the later regression stage, it has been found that the surrounding erythema declines with development of the white streaks. Added to that the vacuolated cells transform across the stages to xanthomatised cells, that it become yellowish that can be distinguished across the three stages. However, the branched and linear vessels are constantly appreciated in the three stages as well [4-6].

The main observed dermatoscopic feature of JXG is the setting sun appearance which can be appreciated in the three stages [4]. Moreover, clouds of pale yellowish globules can be seen and represents lipid laden histiocytes in the upper dermis, with subtle pigment network and white streak which implies fibrosis process [4]. Dermoscopy is valuable to differentiate between different stages of JXG and other similar conditions of yellowish papules and nodules [6].

Cutaneous horns are seldom reported in children, and histopathologic examination remains to be the only way to establish the diagnosis of JXG. There are however few reports cases of JXG in the existing literature of cutaneous horns in children.

JXG constitutes a buildup of histiocytes that devoid of birbeck granules (non-Langerhans cells) that can be made distinctive by special stains [5].

Histology is mandatory in most cases to establish a confirmed diagnosis of dermatoscopic histologic correlation; nonetheless this cannot possible in all cases, especially when the lesion follow the natural call for regression and dissolution [7].

A careful dermatoscopic examination in hand with the clinical picture should be exercised to avoid unnecessary surgical intervention in infants and young children. Following up with a dermatoscope will expand the physician knowledge to the different stages of the JXG evolution. Thus the extended application of dermatoscope in pediatric age group will be of paramount to aid in diagnosis and avoid unnecessary painful surgical excision.

To the best of my knowledge visceral lesions have not been described in this young girl.

References