Case No. 25

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Patient: A 1-year-old Thai boy from Bangkok

Chief complaint: Multiple small yellow-red papules at face, trunk and upper extremities for 6 months

Present illness:
A 1-year-old boy presented with asymptomatic, multiple yellow-red macules and papules for 6 months. Initially, the lesion started on his face. The number of lesions gradually increased predominantly on head and neck region and spread to the upper trunk and upper extremities. He was otherwise healthy and developmentally normal. Both his weight and height were at the 50th and the 25th percentile, respectively. Previously, he treated at clinic with topical corticosteroids but his symptom didn’t improve. Later, macules turned into papules and were spreading to the lower trunk, lower back, upper thighs continuously.

Past history: The patient was born full-term and healthy with no illness or abnormality.

Family history: His parents were not consanguineous and no family member experienced the same condition.

Physical examination:
- GA: active boy, normosthenic build, not pale conjunctiva, no icteric sclera
- Oral cavity: normal mucous membrane, normal teeth
- CVS & RS: normal
- Abdomen: soft, not tender, no hepatosplenomegaly
- Ophthalmologic exam: pending

Dermatological examination:
Multiple discrete, asymptomatic, yellow-red erythematous dome-shaped papules on face, trunk, back and upper thighs

Histopathology: Slide no. 61-2343 from right cheek
Section display a focal aggregates of foamy histiocytes in the upper dermis. Touton giant cell are identified. In addition, some lymphocytes are present within the lesion. No cholesterol cleft is evident.
Diagnosis: Benign cephalic histiocytosis progressing into Juvenile xanthogranuloma

Treatment: Reassure and follow up

Discussion:

Non-Langerhans cell histiocytosis (NLCH), or class II histiocytosis represents a board group of different disorders characterized by the proliferation of histiocytes other than Langerhans cell (LC), including Juvenile xanthogranuloma (JXG), generalized eruptive histiocytosis (GEH), benign cephalic histiocytosis (BCH) and etc. ¹

Benign cephalic histiocytosis (BCH) is an extremely rare form of non-Langerhans cell histiocytosis in which multiple small yellow to red-brown papules. BCH lesions initially develop on the cheeks and forehead, and later involving the whole head, neck and sometimes upper trunk and arms. The mucous membranes, palms, soles, and viscera are not involved, and serum lipid are normal. BCH resolves spontaneously after a mean of 26 months from onset, leaving small atrophic scars. The histologic features predominate of vacuolated histiocytes present in early lesion. Older lesions may contain slightly foamy histiocytes. Touton and foreign body giant cells may also be seen. CD68+ cells strongly support the diagnosis. ²

Juvenile xanthogranulomatosis³,⁴, asymmetrically distributed, scatter lesions, red-brown papules predominantly on the face, scalp, and upper body that may be more widely extracutaneous involvement can occur, particularly eye manifestation, although unusual(seen in approximately 0.4% of patients, with infants younger than 2 years, and multiple skin lesions being at greatest risk) is associated with significant complications including glaucoma, hyphema, uveitis, iritis, heterochromia, proptosis, and occasionally blindness, and therefore these infant should be followed by an ophthalmologist. Rarely other organs could be involved, including central nervous system, bones, lungs, liver, spleen, gastrointestinal tract, pericardium, and soft tissues. The normal lipids were consistent with JXG.

The histologic findings⁵ of vacuolated histiocytes together with lipid laden foamy histiocytes, and Touton giant cells were seen. The exact histologic features seen are dependent on the age of lesion biopsied, with the latter occurring in mature lesions. BCH and JXG are known as non-Langerhans cells histiocytosis that share clinicopathological and immunohistochemical characteristics.
In our case showed multiple yellow-red macules and papules 6 months ago. Initially the lesions confined to head and neck region. Then they increased and spreaded to his upper trunk and upper extremities. Therefore, we thought BCH is the diagnosis.

After several follow-ups the new lesions presented on the lower trunk and upper thighs. Differential diagnosis include BCH, JXG, Langerhans cell histiocytosis (LCH). A close histologic relationship and presence of overlapping symptoms observed among BCH and JXG indicate that these entities fall into a spectrum of the same disorder. The histology of JXG findings of vacuolated histiocytes together with lipid laden foamy histiocytes. However, the presence of a uniform infiltrate of large foamy histiocytes readily distinguishes xanthomas from BCH. S100- and CDla negative on special staining is helpful to differentiate from LCH.

There is significant clinical overlap between some of the non-Langerhans cell histiocytosis. Additionally there are reports of BCH progressing into JXG. There is might be the same condition of our patient.

The treatment for BCH and JXG is spontaneous resolution so reassuring for parents are very important. However extracutaneous involvement in BCH and JXG can occur therefore the patient should be followed.

Reference