Case report:

Juvenile xanthogranuloma

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Abstract

Juvenile xanthogranuloma is a rare disorder which may be present at birth, or appears in infancy. It can also occur in adults of all ages, appears with lesions that may be solitary or multiple nodules several millimeters in diameter. The predilection sites are head and neck, but it may occur on the extremities and trunk also. Internal organs such as lung, kidney, gastrointestinal tract etc. can also be involved. The most frequent extra cutaneous location is in the eye. Here we present a case report of a 5-month-old male child with bronchopneumonia with RDS and left upper eyelid papular swelling with ulceration.

Introduction

Juvenile xanthogranuloma (JXG) is a rare disorder belongs to the group of the non-Langerhans cell group of histiocytic proliferative disorder(1). Juvenile xanthogranuloma was first reported by Adamson(2) as single or multiple cutaneous nodules in infancy as congenital xanthoma multiplex. Juvenile xanthogranuloma is present at birth in 5%-17% of cases but it mostly arises during the first year of life(3). A solitary cutaneous lesion is the most common presentation but it may occur as a soft tissue lesion with or without organ involvement(1). Skin lesions are self-limiting and vary in size, they are reddish or yellowish benign papules or nodules which do not require treatment(4,5). Although the head, neck and trunk are the most common sites for JXG, it can appear anywhere on the body, including the groin, scrotum, penis, clitoris, eyelid, toenail, palms, soles and lips(3). The eye, particularly the uveal tract, is the most frequent site of the extracutaneous involvement(6). 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).

Case report

A 5-month-old male child first issue of non-consanguineous marriage was admitted in our hospital with high grade fever, cough, dyspnoea and a small papule of size 2x3 mm on left upper eyelid just lateral to the medial canthus, his RDS scoring was grade-II. He was investigated with: Hb-12.5 gm%, TLC-18500/cumm, P-72%, L-28%, CRP- Positive, chest x-ray- bronchopneumonia, with hepatosplenomegaly on USG abdomen. USG left eye-normal. He was treated with IV fluids and IV antibiotics, discharged on 5th day with good recovery from bronchopneumonia with RDS. Swelling over the left eyelid gradually increased in next one month. The swelling was pale, grayish nodular measuring 8x6x2 mm with central ulceration, he was referred to ophthalmologist, investigated with Intraocular pressure right eye-14 mm Hg, left eye-15 mm Hg, both eye corneal diameters were normal,
anterior chamber fundoscopy- normal, neuro sonography- normal. Local application of antibiotic with steroid ointment was tried but swelling size increased hence excision biopsy done.

**Excision biopsy-** report showed clusters of moderately large, round to polyhedral cells, with pale eosinophilic; foamy cytoplasm & indented or grooved nuclei. Interspersed are small vascular channels. Few multinucleate giant cells seen. Patient is followed regularly for last 8 months with no recurrence of the lesion anywhere on the body.

**Discussion**

JXG is a benign cutaneous fibrohistiocytic lesion and a type of granulomatous process(8). Even though the etiology is unknown, it is believed to result from disordered macrophage response to the nonspecific injury. The main clinical feature is papulonodular lesion, tan-orange in colour and several millimeters in diameter, which may be single or multiple. The frequent sites of occurrence are the skin of head and neck, but may occur on the trunk or extremities also. The eye is the most common extracutaneous site of JXG. Ocular involvement may occur without concomitant skin involvement. JXG was reported by H. G. Adamson in 1905(9) and named in 1954 describing the appearance of cells under microscope(9) In about 10% of cases cases may be present at birth, but mainly affects infant and small children. According to Zimmerman 64% of cutaneous lesions are by age of 7 months and 85% before 1 year(6). The appearance of multiple lesions is common in children younger than 6 months(8). JXG may occur in adults of all ages too, but this onset is infrequent and tend to be more complicated(10). The male to female ratio of cutaneous JXG is about 1.4:1 in children, while in adults no sex predilection exists(10). The diagnosis of JXG is mainly based on characteristic clinical features. The differential diagnosis includes spitz nevi, mastocytomas and dermatofibromas(1). The confirmation of diagnosis can be made by skin biopsy. Histological findings in JXG are dense dermal histiocytic infiltrate and Touton giant cells which are multinucleated, with homogeneous eosinophilic cytoplasmic center and xanthomatization in the periphery(7). Immunohistochemistry has an important role in the differential diagnosis between Langerhans cell histiocytosis(LCH) and JXG(8). JXG lesions usually label strongly with markers CD68, factor XIIIa and often anti CD4(11,12). S-100 protein reactivity, which is marker for the diagnosis of LCH, is typically absent(11,12). In most cases S-100 protein was nonreactive but scattered cells may show weak cytoplasmic reactivity, unlike the more diffuse and intense reaction of Langerhans cells(13). According to Dehner, neither factor XIIIa negativity, nor S-100 positivity should preclude the diagnosis of JXG.

**Conclusion**

JXG is self limiting disorder, skin lesions gradually increasing in size causes esthetic deformity with at a time bedding and secondary infection. In our case
A lesion was involving left upper eyelid and increasing in size causing ptosis with central ulceration leading to secondary infection, hence excision biopsy done without recurrence, still requires regular follow-up for extra cutaneous involvement or recurrence at the site of excision, bronchopneumonia and RDS were presenting symptoms probably not related to JXG.

References: