CASE REPORT

PAPULAR HISTIOCYTOSIS OF HEAD: TYPE OF NON-LANGERHANS CELLS HISTIOCYTOSIS
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ABSTRACT: Papular histiocytosis of the head also known as benign cephalic histiocytosis is a rare, self-limiting histiocytosis that typically starts in early childhood.¹ Erythematous macules, papules and nodules develop on cheeks and spread to the forehead, earlobes and neck. Lesions are asymptomatic, gradually become reddish brown and may spread to involve the trunk, shoulders and back region including buttocks. There is no mucous membrane involvement. We report a case of 1 and half year boy, born of non-consanguineous marriage presented with multiple, asymptomatic, erythematous, yellow to red color papules on cheeks and back. Lesion was slowly progressive in nature for the past 7 months. Clinical and histological features were consistent with papular histiocytosis of head.

KEYWORDS: Papular histiocytosis of head.

INTRODUCTION: Papular histiocytosis of head is a rare form of non-langerhans cell histiocytosis. These are broad group of disorders characterised by the proliferation of histiocytes involving cells other than langerhans cells. No sex predisposition has been reported. The average age of onset is 15 months (Range 2- 66 months). Most cases are in infants and run a benign self-healing course lasting a few years. Most common sites of involvement are the forehead, cheeks, trunk, and skin folds, with mucosal lesions rare. The number of lesions varies from 2 to more than 100 and new ones continue to appear. Primary histopathologic feature shows the presence of a diffuse infiltrate of histiocytes in the upper dermis with close apposition at the dermoepidermal junction. Cells have intracytoplasmic comma shaped bodies,² dense bodies and coated vesicles.

CASE REPORT:
- A 1 and half year old boy, born of non-consanguineous marriage presented with multiple, asymptomatic, erythematous, yellow to red colour papular lesions on cheek, shoulder region and back.
- Lesions were slowly progressive in nature for the past 7 months.
- Lesions were 1st observed over face and gradually progressed to involve upper back and shoulder region.

History and Examination:
- Family history –unremarkable.
- Non-consanguineous marriage.
- Mucous membrane, palm, soles, teeth-normal.
- Papular, coloured lesions present over face, shoulder and back.
- No organomegaly or lymphadenopathy.
- Neurodevelopmental and opthal examination-normal.
DIFFERENTIAL DIAGNOSIS:

- PLANE WART - pt was initially treated for 2 months (Phenolisation was done) no improvement.
- MOLLUSCUM CONTAGIOSUM.
- JUVENILE XANTHOGRANULOMA.
- XANTHOMA DISSEMINATUM.
- PAPULAR MASTOCYTOSIS.
- MULTIPLE SPITZ NEVI.
INVESTIGATIONS:
- ROUTINE BLOOD EXAMINATION-NORMAL.
- LIPID SCREENING-HDL, LDL, CHOLESTROL-NORMAL.
- SYPHILIS SEROLOGY-NEGATIVE.
- X-RAY SCREENING –NO BONY DEFECT.
- PUCH BIOZY WAS TAKEN FROM BACK LESION.

HISTOPATHOLOGICAL RESULT: Biopsy specimen showed normal epidermis and sufficient proliferation of large, pleomorphic epitheliod histiocytic cell with large cytoplasm within the upper and mid dermis without epidermotropism.

DISCUSSION: Papular histiocytosis of the head also known as benign cephalic histiocytosis is a rare, benign non-langerhans cell histiocytic disorder of unknown etiology occurring in infancy and early childhood, and was 1st described by “gianotti et al” in 1971. This disorder is characterized by multiple, yellow-red to brown popular (1-8mm) self-healing lesions occurring initially in forehead, cheek and neck region. Lesion may subsequently extend to upper and lower extremity and buttocks. Mucous membranes, palm, soles and visceral organs are not involved in this disease. The lesions start appearing at 2months to 66 months (avg-15 months). In approximately 50% of cases lesions start appearing at 6 months of age. Males and Females are equally affected. Spontaneous regression of lesions begins on an average b/w 6 to 48 months and complete regression occurs around 50 months leaving hyper pigmented macules but no scars.

Although no systemic disease has been associated with bch there have been 2 reports of DI/DM occurrence with the disease Consistent with the above text, our pt had multiple asymptomatic, yellow-red coloured, papular lesions over head, neck, shoulder and upper back region. Lesions started at the age of 6 mths and were slowly progressive in nature. Palm, soles, and mucous membrane involvement was absent. Histopathological features of BCH show well circumscribed histiocytic infiltrate in superficial to mid reticular dermis. There are also inflammatory infiltrate composed of lymphocytes and rarely eosinophilis. Touton giant cells and foam cells are usually not present. Electron microscopy shows comma shaped or worm shaped bodies in cytoplasm of 5-30% of histiocytes. In contrast with langerhans cell histiocytosis birbecks granules are absent. Immuno-histochemical staining of lesional cells demonstrates positive staining for macrophage/histiocytic markers including factor XIIIa and CD68 and remain negative for langerhan cell markers such as CD1a and S100 protein. Pts biopsy specimen was consistent with the above histopathological features. Immunohistochemical staining was not done due to lack of sources.
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CONCLUSION/TREATMENT: Clinical and histopathological features of lesion were consistent were PAPULAR HISTIOCYTOSIS OF HEAD. Spontaneous regression of lesion occurs months to years after the onset. Regression of lesion starts characteristically where papules 1st appeared. Papules begin to flatten and after a short period of hyperpigmentation, the lesions disappear without leaving scars.

REFERENCES:

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