PAEDIATRIC DERMATOLOGY

AN EPIDEMIOLOGICAL STUDY OF THE ENIGMATIC DISEASE- LANGERHANS CELL HISTIOCYTOSIS (LCH) - REVIEW OF CASES IN 6 YEARS

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BACKGROUND: In 1868 Paul Langerhans(medical student) identified Langerhans cell. Clonal proliferation of Langerhans cells named as Langerhans cell histiocytosis (LCH),rare but presents as chronic dermatitis.LCH remains enigmatic with the issue that it's a neoplastic or reactive disorder. Here we analyse LCH retrospectively over a span of 7 years.

OBJECTIVES: To know the demographic characteristics in terms of age,sex distribution,presentation,complications & prognosis of LCH in pediatric age group.

METHODOLOGY: All data,photograph,treatment & follow up record of 6 years(Jan 2010 to Jan 2016.) regarding all LCH patients was collected & analyzed retrospectively. Being a rare disease the study population was small.

RESULTS: Total 12 cases(7M & 5F)were confirmly diagnosed based on clinical features, histopathology, and immunohistochemistry(CD1a & S100).75% were within 2.5 years of age. Most common cutaneous presentations were seborrheicdermatitis(33.3%) followed by generalizedeczema(25%),vesico-pustules(8.3%),miliaria(18.3%)etc.Bone lesion was commonest among extracutaneous manifestations (83.3%).One patient with lung, bone, lymphoreticular involvement was declared cure & had no recurrence in 3 years follow-up & One died who presented with seborrheic dermatitis,developmental delay & developed generalized edema, exophthalmos, hemoptysis.One male patient presented with forehead swelling & exophthalmos later developed miliarial lesions.

CONCLUSION: Seventy five percent cases were within 2.5 years with bit male preponderance.Early age of onset,exophthalmos,multi organ involvemeccnt was having a bit poor prognosis. Limited cutaneous disease & aggressive chemotherapy from the very beginning favors remission & less recurrence. The prognosis was not as bad as it was thought.