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Permanent Consequences in Langerhans Cell Histiocytosis Single Center Study

Arjmandi Rafsanjani Kh., Bahoush GR., Vossugh P., Hedayatiasl AA., Faranoush M.

Abstract:

Introduction: Langerhans cell Histiocytosis (LCH) is a rare disease characterized by clonal prolifera-tion of Histiocytosis in different tissues. Permanent consequences (PC) described among subjects with Langerhans cell histiocytosis (LCH).In this study we report the prevalence of permanent sequel among long – term survivors of LCH in our center. Methods: We had 30 cases of LCH from 1989 – 2001 who came for at least 3 years after diagnosis. In-formation has been collected from their disease history, and on type and date of onset of any PC. The cumulative risks of developing a PC have been calculated from the date of LCH diagnosis using the Kaplan-Meier and non-parametric method. Results: Among 30 patients 53.3% were female, 46.67% male, mean age at diagnosis 56.86%±7.79 months (range 7-156), median 42.5 months. 19 (63.33%) had single system (SS) and 11 (36.66%) mul-tisystem (MS). Mean age at SS 5.97 ±1.03 yr, mean age of MS 7.59±1.05 yr. Mean age at follow up 11.3 ± 0.9 yr, median 11.5 yr, range 4.16-22 yr. Mean duration of follow up 6.57 ± 0.76 yr, median 5 yr, and range 3-18 yr. Nine of 30 cases (30%) had at least 1PC; in SS (26.3 %) and in MS (36.7%). The most frequent PC was diabetes insipidus (DI) 16.7%, in SS 5.26% , in MS 36.36%, the difference is significant $P<0.05$.Orthopedic abnormalities 10% which was only in SS (15.79 %), growth retardation (GR) 13.34%. Conclusion: The prevalence of PC in our patients is low which could be due to a small sample and on the other hand as most of our patients had single system involvement, the exact prevalence of PC is not clear. Analysis of cumulative risk shows that some types of PC may become manifest many years from diagnosis and long term follow up is necessary for all patients.

Keywords:

LCH, long – term survivors ، DI and other problems

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