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abstract

Pediatric Langerhans Cell Histiocytosis – Clinical Profile and Outcomes at a Tertiary Center in Chennai, Tamilnadu: A 10 Year Experience

Madhulika Chittala, Meena Sivasankaran, Arathi Srinivasan

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abstract

Pediatric Langerhans Cell Histiocytosis – Clinical Profile and Outcomes at a Tertiary Center in Chennai, Tamilnadu: A 10 Year Experience

Authors: Madhulika Chittala, Meena Sivasankaran, Arathi Srinivasan

Affiliation: Kanchi Kamakoti CHILDS Trust Hospital

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Introduction: Langerhans cell histiocytosis (LCH) is a rare and complex disorder that affects children and can manifest in various ways. Recognising these variations is crucial, as they can greatly impact treatment strategies and overall prognosis for those affected. This study aimed to illustrate the diverse clinical profiles and outcomes of children with Langerhans cell histiocytosis.

Methodology: This retrospective study was conducted at the Department of Pediatric Hemato-oncology, where we reviewed the medical records of children under 18 years old diagnosed with LCH from January 2014 to January 2024.

Results: A total of 45 children were analyzed, and 40 were included in our study. The majority of patients were males (62.2%) compared to females (37.8%), with a median age of 18 months (IQR: 12 – 78 months), reflecting a young patient cohort with wide variability. Most frequent presentations were seborrheic dermatitis of scalp 13(28.9%), organomegaly 11(24.4%), cervical lymphadenopathy 8(17.8%), scalp swelling 6(13.3%), abdominal distension 5(11.1%), CNS risk lesions, skin lesion 4(10%) each, limping gait, diabetes insipidus 3(6.7%) each, anaemia, nail changes 2(4.4%) each and portal hypertension, deranged liver enzyme, microcephaly were each 1(2.2%) respectively. All cases were biopsy-proven and were CD1a (100%) and S100 (97.5%) positive.

Among the 40 children, 18(45%) were diagnosed as Singlesystem LCH, in which 6 had multifocal bone, 4 had unifocal bone, 1 had special site involvement, respectively, and 22(55%) were diagnosed as Multisystem LCH, with 40% Risk Organ involvement. Overall organ involvement in Singlesystem LCH and MultisystemRO+ was Bone (72.2%) and Skin (62.5%), Liver (31.3%), respectively, which shows that the most commonly affected by the disease. Skin-only involvement was seen in 4(10%) children, and liver-only involvement was seen in 3(7.5%) children. The children were treated according to Histiocyte Society treatment guidelines, LCHIII protocol for 25(62.5%) and LCHIV protocol for 7(17.5%) children, with further therapy depending on response.

At the end of the initial phase assessment, no active disease was achieved by 18 children (45%), active disease-progression in 2(5%) children, and active disease-intermediate risk in 9(22.5%) children. Out of the 11 children with active disease, 10(25%) children went for salvage chemotherapy, and 1 child opted out. The outcome of our study showed that

79.4% of children had completed resolution of the disease, and 17.6% had recurrence of the lesion during follow-up.BRAFV600E mutation was positive in 4 children, 4 children were assigned only to follow up, 3 children had died due to disease progression, and 1 child had undergone curative liver transplantation, given liver LCH with decompensation. Overall survival was 90.6%, and overall EFS was 94.4% with a median follow-up period of 28 months.

Conclusion: LCH is a rare disease, and most related publications tend to be multi-institutional. This report was conducted based on the past decade, including 40 pediatric patients with adequate follow-up duration. Similar to earlier reports, the demographics of LCH in our study showed early disease onset and a male predominance. The organs most affected by disease in our study were the bone, skin, and liver, which aligns with previously reported cases in the literature. The presence of BRAF-V600E in circulating blood has been linked to the recurrence of disease.

In our study, 4 children were BRAF-V600E positive, with 2 children on Trematinib. In our study, patients with SS disease and MS-RO- exhibited excellent survival rates, and the recorded deaths occurred in the MS-RO+ group. Our findings support the hypothesis that organ involvement is a significant negative predictor of outcomes in patients with LCH. Our study highlights the need to stratify patients into risk groups to guide clinical decision-making and treatment protocols. The SS group shows better outcomes and fewer risk organ involvements, while the MS group exhibits significant clinical challenges and poorer survival rates. Targeted therapies and tailored approaches are recommended for high-risk groups.