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ELEVENTH ANNUAL MEETING

A RARE, CHALLENGING DISEASE ; LANGERHANS CELL HISTIOCYTOSIS - A SINGLE CENTER EXPERIENCE

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OBJECTIVE

Langerhans cell histiocytosis (LCH) is a rare, clonal disorder derived from CD1a-positive and CD207-positive immature myeloid dendritic cells , with a wide range of clinical presentations. Diagnosis of LCH is difficult and may be delayed due to its rarity and wide clinical manifestation.

METHOD

In this study ;we aimed to retrospectively analyse our patients with LCH in terms of diagnostic procedure ,clinical course and treatment outcomes for the last 12 years.



RESULTS

Twenty-nine patients who were diagnosed with LCH and received treatment between January 2010 and September 2022 at Ege University Hospital (İzmir, Türkiye) were included . Overall, 18 patients were male (62%), with a male-to-female ratio of 1.6:1. The median age at diagnosis was 36 years (range,17-62 years). The median time from disease onset to diagnosis was 3 months (range 1–180months). After a median follow-up of 38-months (range 3–153 months), all patients are alive.

At the time of diagnosis, 18 patients were classified as unifocal single system LCH (62,1%), 2 patients had pulmonary single system LCH (6,9%), 2 patients had multifocal single system (6,9%), and 9 patients had multisystem LCH (31%).

Among the 18 patients with unifocal single system or organ , 14 had unifocal bone lesions, 2 had isolated pulmonary system, 1 had ear involvement and 1 had other single lesions (tooth). The most prevalent involvement in multisystem patients was the pituitary gland (66%), followed by the bone (55%), lung (55%), and vulva (33%). Ten (34,5%) patients had risky organ involvement.

Seven patients (24,1%) had received systemic chemoterapy , 11 (37,9%) had radiotherapy and 5(17,2%)had both chemotherapy and radiotherapy as treatment. Therapies in this study were vinblastine and prednisone-based chemotherapy, cladribine monotherapy , lenalidomide , vemurafenib and radiotherapy . The ORR was 93%, including 23 patients (79,3%) classified as having CR and 5 patients (17,2%) classified as having PR. One patient (3,4%) were evaluated as SD. Three patients had reactivation of disease . Notably, 1 patient was diagnosed with LCH after 15 years and developed central diabetes insipidus.

CONCLUSIONS

Langerhans cell histiocytosis is a heterogeneous disease with various clinical presentations and outcomes. A systemic evaluation is crucial to determine the extent of organ involvement and to guide treatment decisions. Individualized and multidisciplinary treatment approaches are necessary for favorable outcomes. Adult-onset LCH, although rare, should be considered in cases of central diabetes insipidus and spontaneous pneumothorax. While LCH is generally considered a benign disorder, it can lead to complications affecting vital organs

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