Experience with Rosai-Dorfman Disease at a National Referral Center in Mexico

M. S. RIVAS-VERA[1], J. ARAUJO-CARDENAS[1] and C. MEDRANO-BORROMEO[2]

[1] Instituto Nacional de Cancerologia (INCan), Mexico City, Mexico
[2] Instituto Mexicano del Seguro Social (IMSS), Mexico

INTRODUCTION

Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis characterized by a highly variable clinical course, ranging from isolated nodal disease to multisystem involvement requiring local and systemic treatment. Institutional data describing adult patients remain limited, particularly in Latin America.

AIM

To describe clinical characteristics, treatment patterns, and outcomes of adult patients with RDD treated at a national oncology referral center.

METHOD

STUDY DESIGN

Retrospective, observational, single-center and descriptive study.

POPULATION

Adult patients (≥18 years) diagnosed with Langerhans cell histiocytosis (LCH) or other non-Langerhans histiocytosis treated at our national cancer referral center.

INCLUSION CRITERIA
- Age ≥ 18 years

EXCLUSION CRITERIA
- None

STUDY PERIOD

January 2014 to December 2024

Total patients identified:

7

patients over a 6-year period, highlighting the rarity of this disease in routine clinical practice.

RESULTS

PATIENT CHARACTERISTICS

Median age at diagnosis
39 years
(range, 26–49)

71.4%
were female

Performance status
ECOG 0–1
in all patients

SITES OF INVOLVEMENT*

Lymph nodes 5/7 (71.4%)
Skin 4/7 (57.1%)
Bone 1/7 (14.3%)
Breast 1/7 (14.3%)

Bone marrow involvement: **2/7 (28.6%)** patients

Fig. 1. Distribution of involved sites in the cohort.

IMMUNOHISTOCHEMISTRY

S100	Positive in 6/7 (85.7%)
CD1a	Negative or unavailable
CD207/ Langerin	Negative or unavailable

Immunoprofile supports the diagnosis of **non-Langerhans histiocytosis.**

Fig. 2. Immunohistochemical profile.

TREATMENT APPROACHES*

Radiotherapy (n=5)
Dose: 30–45 Gy
- Complete response: **3/5** (60%)
- Partial response: **1/5** (20%)
- Progressive disease: **1/5** (20%)

First-line systemic therapy (n=5)
Regimens: corticosteroid-based or CHOP
- Variable responses

Three patients (**3/7, 42.9%**) required **second-line treatment.**

Fig. 3. Treatment approaches and responses.

TREATMENT RESPONSE (OVERALL)*

Complete response 3/7 (42.9%)
Partial response 2/7 (28.6%)
Progressive disease 1/7 (14.3%)
Not evaluable 1/7 (14.3%)

Fig. 4. Overall treatment response.

OUTCOMES

Overall survival
100%
at last follow-up

5/7 (71.4%)
patients alive without active disease

2/7 (28.6%)
patients alive with persistent disease

Fig. 5. Outcomes at last follow-up.

DISEASE PRESENTATION / SUBTYPES*

Predominantly nodal-cutaneous phenotype

Nodal involvement	5/7 (71.4%)
Cutaneous involvement	4/7 (57.1%)
Bone marrow involvement	2/7 (28.6%)

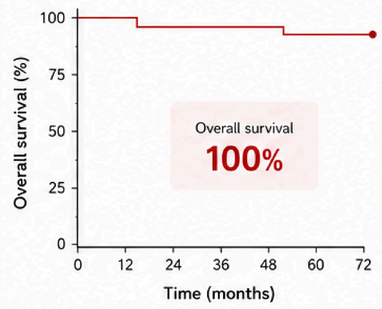
Other extranodal sites
- Bone: 1 patient (14.3%)
- Breast: 1 patient (14.3%)

No true CNS involvement was documented.

RDD showed heterogeneous patterns of involvement, ranging from nodal disease to multisite/extranodal presentations.

Fig. 6. Clinical presentation and subtypes/extensión de la enfermedad.

SURVIVAL SUMMARY



Overall survival **100%**

Median follow-up: **33** months (range, 2–84)

Fig. 7. Kaplan–Meier curve for overall survival.

* Percentages are calculated based on the total cohort (n = 7).

CONCLUSIONS

In this institutional RDD cohort, patients had preserved functional status and predominantly nodal-cutaneous involvement. Clinical presentation and treatment requirements were heterogeneous, reflecting the variable behavior of this rare disease.

Radiotherapy demonstrated favorable local responses, whereas systemic therapy showed variable efficacy and some patients required additional treatment. Despite persistent disease in a subset of patients, overall survival at last follow-up was 100%.

These findings provide real-world evidence on the clinical spectrum and management of RDD in a Mexican referral center and highlight the importance of individualized treatment strategies for this rare histiocytic disorder.

ACKNOWLEDGEMENTS

The authors acknowledge the Hematology Department of the Instituto Nacional de Cancerología (INCan) for its institutional support and contribution to this study.

CONTACT INFORMATION

Janeth Araujo-Cardenas, MD. Hematology Department, Instituto Nacional de Cancerologia (INCan). Mexico City, Mexico
Email: janetharaujo@unam.edu

REFERENCES

1. Emile JF et al. Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages. Blood. 2016;127:2672-2681.
2. Goyal G et al. The Mayo Clinic histiocytosis working group consensus statement. Mayo Clin Proc. 2019;94:2054-2071.
3. Durham BH et al. Activating mutations in CSF1R and MAPK pathway genes in histiocytic neoplasms. Nat Med. 2019;25:1839-1842.
4. Allen CE et al. Langerhans-cell histiocytosis. N Engl J Med. 2018;379:856-868.
5. Candelaria M et al. Rare histiocytic disorders in Latin America: challenges and perspectives. Hematology. 2020;25:1-7.