



# AN OLD DRUG FOR A RARE HISTIOCYTOSIS:

## Thalidomide as a Pragmatic Option for Extensive Extranodal Rosai-Dorfman Disease in a Resource-Limited Setting

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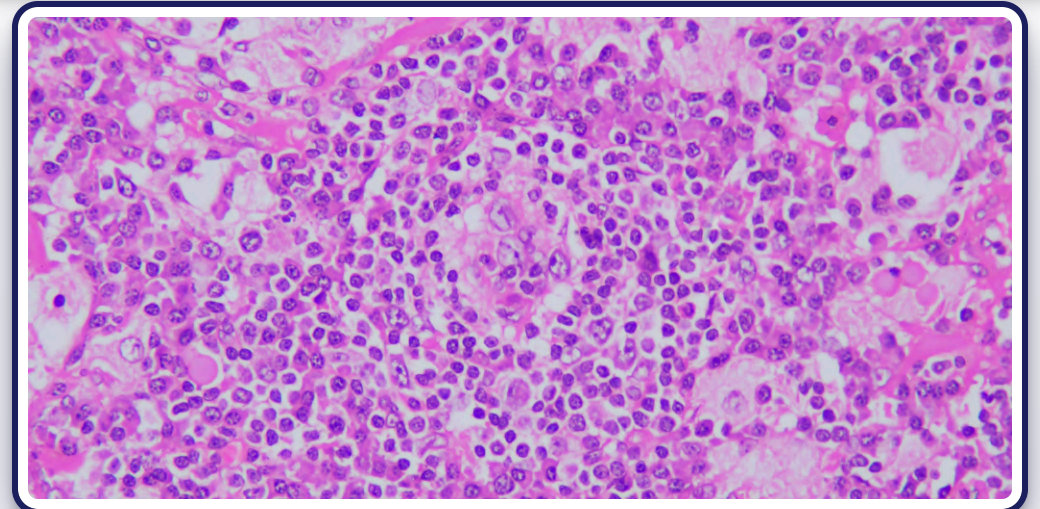
### ● BACKGROUND

- Rosai-Dorfman disease (RDD) is a rare non-Langerhans histiocytosis, about 1 in 200,000.
- It mimics infection and autoimmune disease, so diagnosis is often delayed, especially in TB-endemic regions.
- No trials exist; treatment follows disease extent and symptoms. Targeted agents are unaffordable in low-resource settings.
- **We report steroid-refractory nodal and sinonasal RDD in remission on low-cost thalidomide for 24 months.**

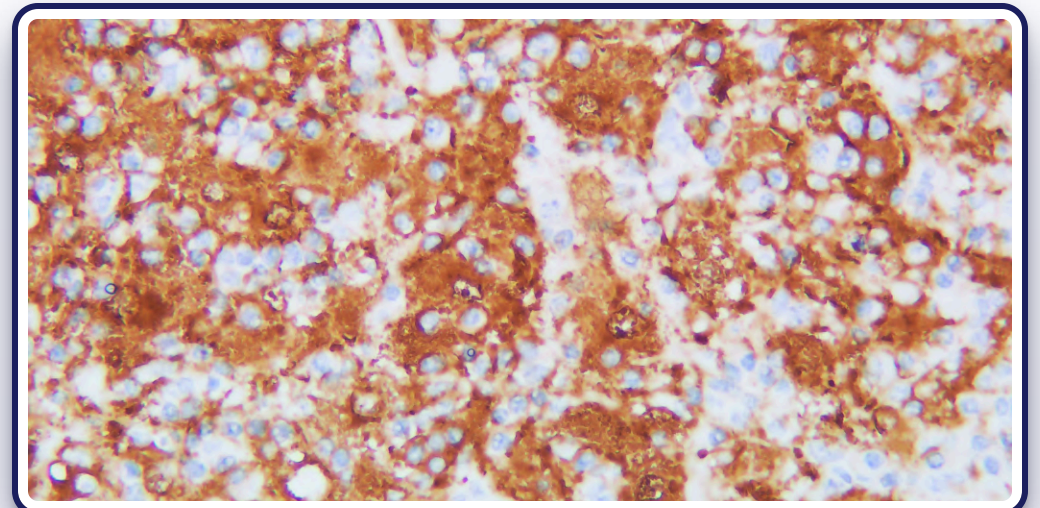
### ● CASE HISTORY

- **Patient:** 51-year-old woman with ischemic heart disease on aspirin + clopidogrel.
- **Presentation:** dry mouth and eyes, bilateral nasal masses with obstruction, then cervical lymphadenopathy, dysphagia, epistaxis, night sweats, arthralgia.
- **Work-up:** Hb 9.1 g/dL, ESR 84 mm/hr; RF, ANA, ENA negative. CT: paranasal sinus mucosal thickening. Ultrasound: "necrotic" nodes, TB suspected.
- **Excluded:** Sjögren's (serology negative) and TB (no granulomas or caseation, GeneXpert negative).
- **Diagnosis:** nasal biopsy read as "inflammatory polyp". Node excision showed RDD; GeneXpert negative.
- **Treatment:** prednisolone + thalidomide 200 mg (Aug 2024). Steroid-free since Feb 2025; thalidomide now 100 mg.
- **Safety:** dual antiplatelet therapy continued throughout because of thrombotic risk with IHD.
- **Outcome:** sicca symptoms resolved, nodes markedly smaller, **ESR 84 → 50 mm/hr**, **Hb 9.1 → 12.9 g/dL**.

### ● MICROSCOPY



**Fig 1.** Emperipolesis



**Fig 2.** S100 staining of histiocytes

- **Emperipolesis:** intact lymphocytes inside large pale histiocytes.

### ● CONCLUSION

**Thalidomide gave durable control of steroid-refractory extranodal RDD, sustained on a reduced dose over 24 months.**

- Affordable second-line option where targeted therapy is out of reach.
- Monitor **thrombotic risk**, **neuropathy** and teratogenicity.
- **Pearl:** look for emperipolesis in "inflammatory polyp" biopsies.
- **Pitfall:** cystic-looking nodes on ultrasound mimic TB; biopsy first.
- Limits: no molecular tests or IgG4; no repeat imaging.

### ● REFERENCES

1. Rosai J, Dorfman RF. Arch Pathol. 1969;87(1):63-70.
2. Mahzoni P, et al. J Res Med Sci. 2012;17(3):304-7.
3. Ablal O, et al. Blood. 2018;131(26):2877-90.