

Skin Rash

[Dermatologist Examining Rash](#)

In multifocal disease, scaly erythematous lesions and red papules can be present and are more pronounced in intertriginous areas. Up to 80% of LCH patients have extensive eruptions on the scalp.

Recurrent Otitis Media

[Recurring Oats-out-of-ear](#)

Due to skull involvement, the mastoid process of the temporal bone is also affected. Due to the destruction of the mastoid antral cells, recurrent otitis media is common.

Diagnosis

Birbeck Granules

[Barbed-wire and Granny](#)

Birbeck granules are collections of rod- or racket-shaped organelles found in Langerhans cells. The exact function of these organelles is not known.

"Tennis Racket" Appearance

[Tennis Racket](#)

Birbeck granules are often said to have a "tennis racket" appearance with a loop and handle.

S100 Positive

[S100-sign](#)

S100 is a cytoplasmic protein that is used as a tumor marker on immunohistochemical staining, particularly for cells derived from the neural crest. CD1a is a marker of antigen-presenting cells.

Management

Observation

[Observatory](#)

If the disease is unifocal and spares the skull without any major systemic manifestations, the patient is kept under observation.

Prednisone +/- Vinblastine

[Predator and Van-blasting](#)

If the patient has skull involvement, multiple bone lesions, or systemic manifestations, immunosuppression with prednisone is initiated. Bone lesions are excised. Chemotherapy with vinblastine is added in systemic disease. Pituitary hormone replacement is also indicated.