Langerhans Cell Histiocytosis

Langerhans Cell Histiocytosis is a rare disorder caused by clonal proliferation of antigen-presenting cells known as Langerhans cells. The defect lies in antigen presentation. This disease can present with lytic bone lesions, most commonly in the skull, or with systemic manifestations like hypopituitarism, diabetes insipidus, skin rash, and recurrent otitis media. On electron microscopy, Birbeck granules with a "tennis racket" appearance can be seen. On immunohistochemistry, cells will show S100 positivity. Asymptomatic solitary bone lesions in low-risk anatomical regions are managed with observation. Prednisone with or without vinblastine is given for bone lesions in high-risk anatomical regions or with systemic manifestations.



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Characteristics

Antigen Presentation Defect
Ant-gem Presentation Defected
Langerhans cells are antigen-presenting dendritic cells in the epidermis. These dendritic cells migrate from the bone marrow to the skin and lymph nodes. In Langerhans cell histiocytosis, these cells undergo clonal proliferation in the bone marrow.

Lytic Bone Lesions
Punch Through a Light-bulb Bone
In Langerhans cell histiocytosis, infiltration of histiocytes into the bone marrow affects bone architecture. Lytic lesions are present. Bone lesions known as eosinophilic granulomas are present as both unifocal and multifocal lesions. Lytic lesions usually present as bone pain and pathological fractures.

Skull
Skull
The skull is the most common bone to be affected by infiltration. The second most commonly affected bones are the long bones of the upper limb.

Presentation

Hypopituitarism
Hippo-pit-bull-terrier
In multifocal disease, the hypothalamic-pituitary axis is involved and Hand-Schüller-Christian triad is present. Bulging of eyes, diabetes insipidus, and bone breakdown comprises this triad.

Diabetes Insipidus
Dyed-bead-pancreas with Sippy-cup
The triad of diabetes insipidus, exophthalmos, and lytic bone lesions is known as the Hand-Schüller-Christian triad. Diabetes insipidus presents with polyuria due to decreased ADH Secretion.

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

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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.

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