

Langerhans Cell Histiocytosis

Definition

A rare disorder of excess number of histiocytes or macrophages called Langerhans' cells. Although not a true cancer it is usually treated by cancer specialists. Previous names were Histiocytosis X and depending on the signs and symptoms: Letterer-Siwe disease, Hand-Schuller-Christian disease or Eosinophilic granuloma of bone.

Etiology

Unknown

Signs and Symptoms

Several different presentations

- Infants may present with rash (looks like eczema) and large liver and spleen
- Bone pain

Diagnostic Evaluation

- Bone scan
- Ultrasound and CT of abdomen

Staging

No uniform system but outcome depends on the number of organ systems involved.

Treatment

- Since the disease involves an excessive proliferation (number) of histiocytes it responds to anti cancer therapy
- Solitary bone lesions respond to curettage, intralesional prednisone or low dose radiation
- Widespread disease is usually treated with chemotherapy (Prednisone, vinblastine, vincristine, 6MP, methotrexate and etoposide have been used)

Prognosis

Depends on extent of disease but most children are cured.

Links

www.histio.org/society/
www.histio.org/association/index/shtml