LANGERHAN CELLS HISTIOCYTOSIS – A Transition Follow-up Patient

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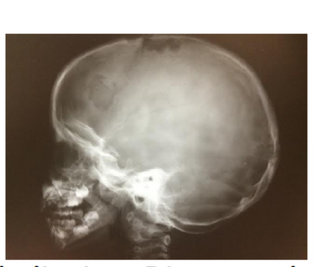
Introduction

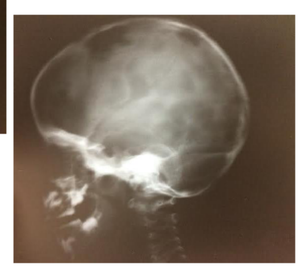
Langerhan Cells Histiocytosis (LCH) is a rare heterogeneous idiopathic clinical entity involving clonal proliferation of Langerhan cells that may infiltrate most commonly bone, skin, lymph nodes or lungs. It affects mainly children between 5 and 15 years. The most common endocrine manifestation is diabetes insipida.

Clinical Case

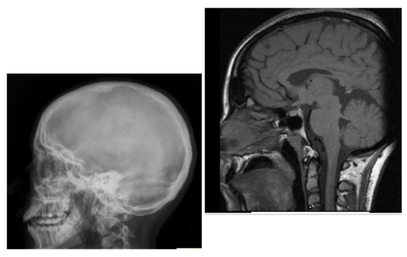
JFM, female, 15 months old
Presenting with a petechial rash, fever, abdominal distension
At physical examination: Retro-auricular petechial lesions, brownish infiltrative lesions over the abdominal, thoracic and back region, abdominal distension with umbilical herniation
Complementary exams: Biochemichally - anemia with thrombocytopenia, a rise of hepatic transaminases. Abdominal ultrassound - hepatomegaly with regular contour and homogenous structure, globous spleen with 12,5cm longitudinally.
Myelogram - Kala-Azar(??)
She began treatment with Glucantime but showed no clinical improvement and evolved to respiratory distress
The myologram was repeated– **immature histiocytes, with no phagocytic content, erythroid hyperplasia. Diagnostic of LCH**
She began chemotherapy, with 6 courses of VP16+PDN (etoposide+ prednisone) with the need of transfusional suport.
Progressive clinical improvement, regression of hepatosplenomegaly and cutaneous lesions were observed.



2 years of age, recurrence with osteolytic lesions
Underwent 9 courses of VP16+PDN



4 years of age, presented with polyuria (up to 7L/day), polydipsia – Diagnosed with Diabetes Insipida. Evidence of new osteolytic lesions.
Underwent 12 curses of VP16+PDN



18 years
Admitted in Transition Follow-up Consult.
Previous medical history: Langerhan Cells Histiocytosis diagnosed 17 years before with 2 recurrences and a total of 27 curses of chemotherapy. Diabetes insipidus as sequelae.
PE: stature 155,3cm, weight 39,9 Kg, BMI 16,5 Kg/m2, Tanner M4P5, no apparent cutaneous lesion, no organo or adenomegaly.
Workup:
-No analytical changes in hemogram, leucogram, biochemical tests or pituitary function; Diabetes Insipida sustained.
- Brain MRI - globous anterior pituitary, neuro-hipophysis not individualized, broadening of pituitary stalk.
- Normal skeletal and thoracic radiographies
- Medicated with: vasopressin 0,1 mg 2id

Discussion

Since it affects mostly children, the majority of published literature about LCH concerns pediatric patients. Our patient reached adulthood and now we face the challenge of planning the future follow-up, concerning the risks of recurrence of the primary disease and the consequences and long-term effects of the treatment of a childhood cancer.

Transition Follow-up requires a retrospective analysis of the past medical history, a comprehensive evaluation of present state and a planification of the future care throughout adult life.

References:
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