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Images in hematology-oncology

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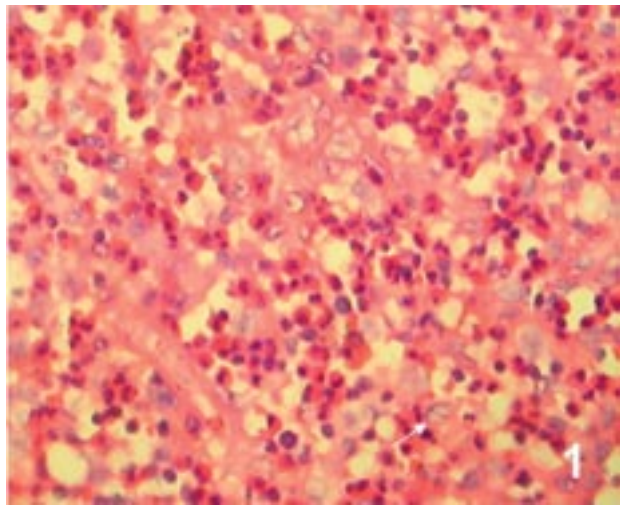
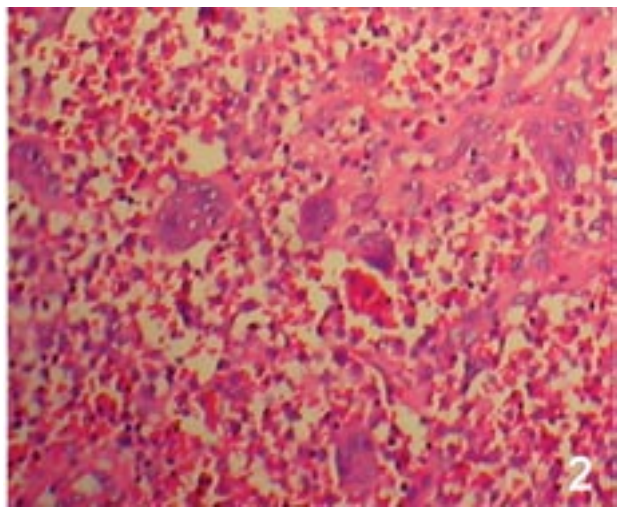
A 6-year-old girl with diabetes insipidus and lytic lesions in the skull

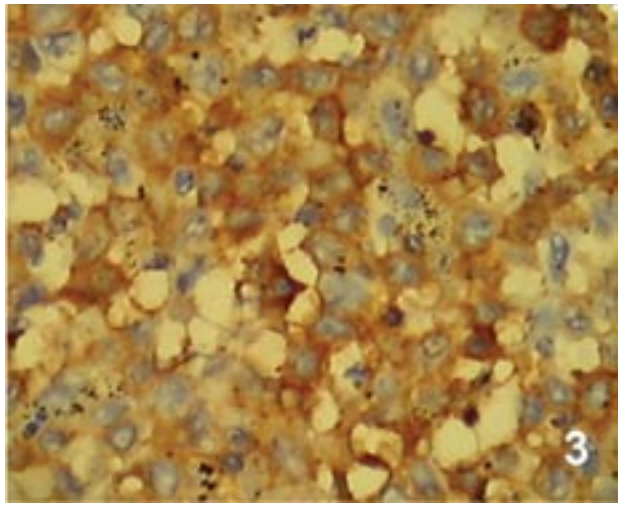
CLINICAL HISTORY

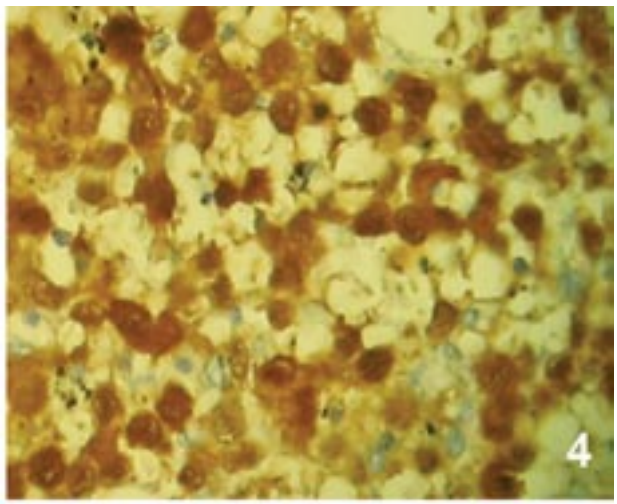
A 6-year-old girl presented with a complaint of softening in her skull. Previously she was diagnosed with diabetes insipidus. In physical examination, a 2 centimeter bone defect was observed. A cranial tomography was performed and three lytic lesions which contain soft tissue component, invading the frontal and the left parietal bone was reported. Abdominal ultrasonography was normal. A surgical excision of the lesion was performed. Microscopic examination of the excision specimen showed hemorrhage and inflammatory-cell rich lesion consisting of large cells with moderate to abundant cytoplasm and eccentrically-located, irregular, kidney-shaped or 'coffee-bean' nuclei with prominent longitudinal grooves (Figure 1). Nuclei also showed folding and indentation. Admixed eosinophils and osteoclast-like multinucleated giant histiocytes were noted (Figure 2). A few granulomas were also observed. Neoplastic cells showed membranous reactivity for CD1a (Figure 3) and cytoplasmic reactivity for S100 (Figure 4).

What is your diagnosis ?









PATHOLOGIC DIAGNOSIS

Langerhans' Cell Histiocytosis

DISCUSSION

Histiocytic syndromes are described as abnormal proliferations of macrophages and histiocyte-like cells (1). Dendritic cells and Langerhans' cells which are characterized by dendritic cytoplasmic processes and the presence of large amounts of cell-surface class II molecules are part of the immune system and are believed to be the most important antigen-presenting cells in the body by some investigators (2). Langerhans' cells are located within the epidermis. Langerhans' Cell Histiocytosis (LCH), also named as Histiocytosis X, makes up Class I Histiocytosis group and covers three disorders: Eosinophilic Granuloma, Hand-Schüller-Christian Disease, Letterer-Siwe Disease (1). It occurs in children and young adults with an estimated prevalence of 1/200000/year. Clinical presentation is variable, ranging from a single location in the bone to severe multivisceral involvement (i.e. lungs, bone marrow, liver, spleen) leading to dysfunction of vital organs due to progressive fibrosis of the tissues involved. Diagnosis is always based on cytological or histological examination. For definitive diagnosis, in addition to morphologic examination membranous expression of CD1a antigen on pathological cells or demonstrating Birbeck granules by electron microscopy is required (2). This case is a typical case of LCH fulfilling all the diagnostic criteria.

References

1- Komp DM, Perry MC. Introduction: The histiocytic syndromes. Semin Oncol 1991;18:1.
2- Robbins Pathologic Basis of Disease. 5th edition; 171-173, 666-667.