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Neurological image

A 27-year old female with a lytic skull lesion

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1. Clinical background

A 27-year old female presented with 2-month-old history of a tender nodule in the left parietal scalp region. She did not have any other neurological symptoms. On exam the lesion was moderately tender and attached to the underlying bone. Her neurological examination was unremarkable. She did not have any systemic signs or symptoms. She underwent MRI of the skull and brain and subsequently underwent craniectomy for resection of this skull lesion (Fig.1).

2. What do you think is the diagnosis of the patient ?

1. Metastatic lesion
2. Lymphoma
3. Osteomyelitis
4. Myeloma
5. Langerhans cell histiocytosis ("histiocytosis X")

1) Answer : The correct choice is 5.

2) Discussion

The MRI shows a lytic lesion involving the left parietal bone measuring $2.9 \times 2.5 \times 1.5$ There is enhancement within the area of lytic change as well as soft tissue extension into the subcutaneous fat with destruction of both the inner and outer tables of the calvarium. There is also enhancement and minimal involvement of the dura. The histopathology of the resected lesion shows clusters of medium to large atypical cells with grooved or lobulated nuclei, and abundant pink cytoplasm. These are accompanied by a mixed inflammatory infiltrate consisting of lymphocyte, neutrophils and eosinophils. Focal osteoclast like giant cells and reactive woven bone formations are seen. Immunocytochemistry shows that many cells are positive for CD1a, a marker consistent with Langerhans cell histiocytosis or histocytosis X.

Langerhans cell histiocytosis is a disease that has three forms : Eosinophilic granuloma (pulmonary or osseous), Hand-Schuller-Christian disease (multiple organs) and Letterer-Siwe's disease, which typically is aggressive and involves abdominal viscera. Eosiniphilc granuloma with dural involvement is very rare.(1) Isolated lesion limited to skull can be effectively treated with surgery. Multiple lesions or viceral involvment may require radiotherapy and chemotherapy, respectively. It is important to note

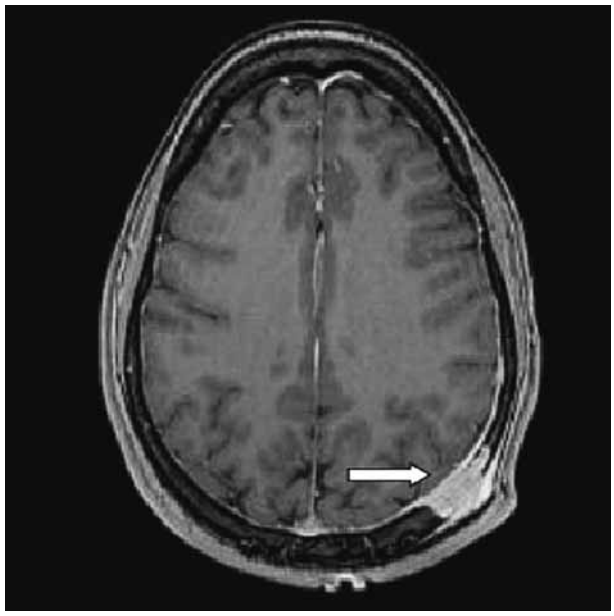

FIG. 1A. — MRI (T1 post contrast) shows a bright lesion involving the posterior parietal skull and the adjacent tissue (arrow).

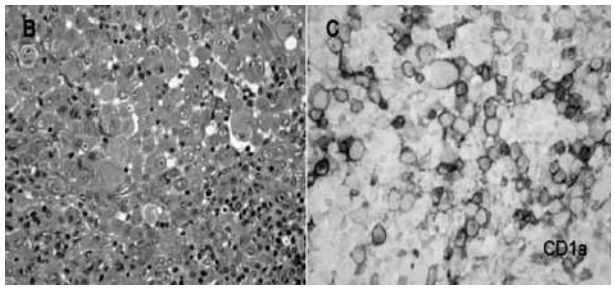

Fig. 1B. — Histopathology of the lesion shows a mixed population of inflammatory cells including many large histiocytes with lobulated nuclei.
Fig. 1C. — Immunocytochemistry shows that many of the histiocytes are positive for CD1a.

that these can have future visceral recurrences. After initial treatment, careful long-term follow up is recommended (2).

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