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An unusual case of intracerebral Non-Langerhans cell histiocytosis with review of the literature

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Aims

In the face of an intracerebral case of Non-Langerhans cell histiocytosis (NLCH), the diagnostic procedure and the clinical handling of this rare histiocytic tumorous lesion are demonstrated and differential diagnoses as Rosai-Dorfman or Erdheim Chester diseases are discussed.

Methods

A stereotactic serial biopsy of frontal brain was taken from a patient with unclear contrast enhancing masses around the sellar region, the hypothalamus, and a wide lining intraventricular. Smear preparations, conventional histology, immunohistochemistry and electron microscopy were done to rule out the etiology of the tumorous lesion.

Results

The morphological analysis of our case revealed a histiocytic proliferation, which, in view of the immunohistochemical and ultrastructural findings, was classified as Non-Langerhans cell histiocytosis. No infectious agents could be detected and no lymphoma-like features could be observed.

Conclusion

This case shows the unusual manifestation of a primary intracerebral Non-Langerhans cell histiocytosis, which begins as a solid mass around the sellar region and shows a remarkable extension as a flat lining of inner and outer liquor spaces. Stereotactic brain biopsy technique today is a valuable high tech tool to clarify intracerebral tumorous lesions with a minimum of risk for the patient and with the possibility to perform all necessary tissue investigations.