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# CT and MR Findings in Diffuse Cerebral Histiocytosis: Case Report

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Intracranial involvement with histiocytosis is usually limited to the hypothalamus and associated with extensive extracranial disease. We report the CT and MR findings in a patient with biopsy-proved histiocytosis presenting as multifocal intracranial involvement without any systemic disease. While extrahypothalamic CNS involvement by histiocytosis is well recognized [1–9], infiltrative brain involvement in the absence of systemic disease is unusual.

## Case Report

The patient is a 19-year-old woman with a history of autoimmune thyroiditis, hypogonadotropic hypogonadism, and pancreatitis who was incidentally noted to have diabetes insipidus. A postcontrast cranial CT study (GE 8800 scanner, General Electric, Milwaukee) revealed a thickened pituitary stalk and innumerable enhancing nodules throughout the cerebral hemispheres and basal ganglia (Figs. 1A and 1B). Little edema and no bone involvement were appreciated on CT. MR imaging (0.5-T Gyrex S5000 scanner, Elscint, Boston) demonstrated large confluent areas of increased signal on both long TR/short TE (1800/27/4) and long TR/long TE (1800/85/4) images (Figs. 1C and 1D). Contrast-enhanced studies were not obtained.

Shortly after the imaging studies were performed the patient developed hallucinations and disorientation associated with dystonic posturing and choreiform movements. An extensive radiologic work-up—including chest X-ray, skeletal survey, and bone scan—was unremarkable. The results of a bone marrow biopsy were normal. Tissue from a frontal lobe biopsy (reviewed at several institutions) showed multicentric intraparenchymal and leptomeningeal differentiated histiocytosis X (Fig. 1E). Despite radiation therapy the patient has developed progressive cognitive impairment with extrapyramidal features.

## Discussion

Histiocytosis is the general term applied to eosinophilic granuloma, Hand-Schüller-Christian disease, and Letterer-Siwe disease, all of which are characterized by abnormal proliferation of large histiocytes containing pathognomonic inclusion bodies in their cytoplasm [1]. While the skeleton is the most commonly affected site, hypothalamic involvement, causing diabetes insipidus, is a well-known complication. Intracranial involvement is thought to be the result of meningeal extension in most instances [5].

Intracranial histiocytosis outside of the hypothalamus is an uncommon occurrence. Lesions have been reported in the cerebral hemispheres, cerebellum, optic chiasm, and spinal cord [1, 3], and are usually associated with hypothalamic and systemic involvement [6].

Rarely, histiocytosis can present as multiple intracranial nodules or, even more rarely, as an isolated hemispheric mass [6]. Some of those patients who present with an isolated hypothalamic mass will go on to develop multiple parenchymal nodules over the ensuing 1–2 years. Whether the lesions are single or multiple, most authors have described contrast-enhancing lesions on CT as being surrounded by edema [4]. Graif and Pennock [2] described the MR findings in three patients in whom hypothalamic and extrahypothalamic CNS lesions were manifest by high signal on T2-weighted studies and were surrounded by edema, similar to our case [2]. However, in all their patients, there was evidence of systemic disease.

In the absence of typical skeletal lesions, the diagnosis of CNS histiocytosis can be suggested only as part of a differential diagnosis. When there is diffuse intraparenchymal involvement that includes the hypothalamus and neurohypophysis, then histiocytosis should be considered a possible cause—albeit a rare one. Thus, in this case, the cerebral and hypothalamic involvement caused us to focus on granulomatous disease (i.e., tuberculosis, sarcoid, etc.), lymphoma, and metastases, with histiocytosis considered somewhat less likely.

Although our patient has not responded to radiation therapy and chemotherapy and continues to deteriorate, other authors have described a good response in similar situations [6, 8]. Histiocytosis limited to the hypothalamus often is successfully treated with radiation therapy. While extrahypothalamic disease is unusual, imaging is important in the delineation of the extent of disease for planning radiation therapy.

In conclusion, the appearance of multifocal hemispheric, basal ganglial, and hypothalamic lesions is nonspecific. While a distinctly unusual manifestation of histiocytosis should be considered in the differential diagnosis, this remains a histologic diagnosis.

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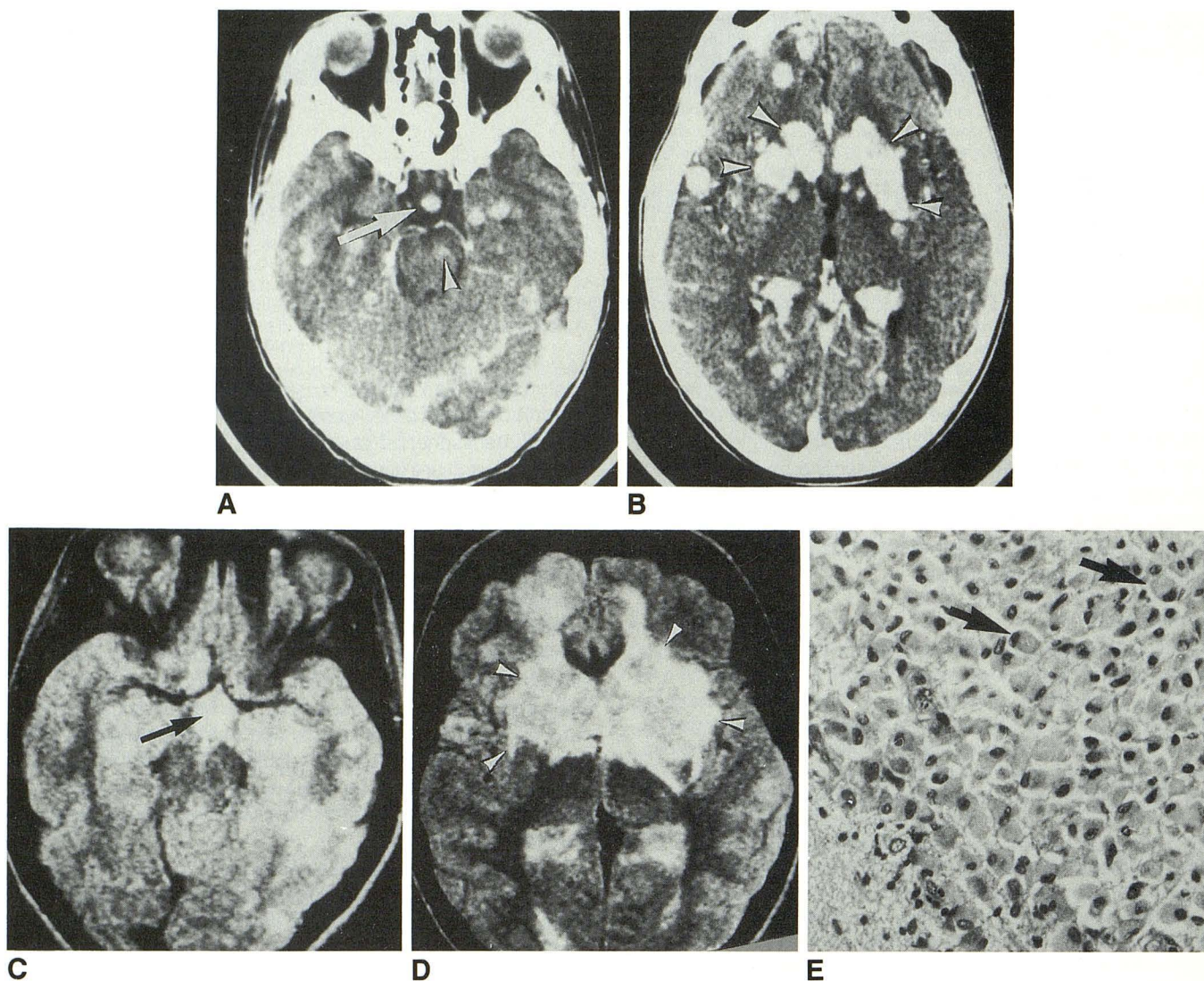


Fig. 1.—19-year-old woman with history of autoimmune thyroiditis, hypogonadotropic hypogonadism, and pancreatitis; incidentally noted to have diabetes insipidus.

A, Contrast-enhanced CT scan through suprasellar region shows marked enlargement and enhancement of infundibulum (arrow), a left anterior basis pontis nodule (arrowhead), and multiple temporal enhancing nodules.

B, Contrast-enhanced CT scan through basal ganglia shows multiple solidly enhancing hemispheric nodules with confluent basal ganglial lesions (arrowheads).

C, Long TR/short TE (1800/27) MR image through suprasellar region shows homogeneous hyperintensity and fullness of suprasellar cistern (arrow); there are scattered areas of parenchymal hyperintensity in the temporal lobes and right posterior gyrus rectus.

D, Long TR/long TE (1800/85) MR image through basal ganglia shows confluent, infiltrative hyperintensity centered in the basal ganglia and extending into the forceps minor (arrowheads).

E, Paraffin section from cerebral biopsy shows infiltration of gliotic neuroparenchyma by large histiocytes (arrows). (H and E,  $\times 145$ ).

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