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AJNR Am J Neuroradiol 1991, 12 (1) 82
http://www.ajnr.org/content/12/1/82.citation

Coexistent Intra- and Extracranial Mass Lesions: An Unusual Manifestation of Histiocytosis X

Eosinophilic granuloma, Hand-Schüller-Christian disease, and Letterer-Siwe disease are three disorders characterized by proliferating histiocytes. Collectively, they are called histiocytosis X. Histiocytosis X often presents with lytic calvarial lesions [1]. Hypothalamic involvement is the most common intracranial manifestation [2]. Solitary histiocytosis X within the cerebral hemispheres, although rare, has been reported [3, 4]. We present a case of solitary eosinophilic granuloma of the temporal lobe with coexistent involvement of the dura, calvaria, and temporalis muscle.

Case Report

A 29-year-old man was found unresponsive. He apparently had fallen and had right-sided facial injuries. After regaining consciousness, he had a generalized seizure en route to the hospital. On arrival, he denied having a headache or focal neurologic symptoms and complained only of generalized weakness and nausea. Physical examination showed a slightly lethargic, afebrile patient with right orbital ecchymosis. No focal neurologic deficits were observed. The patient had had a transphenoidal hypophysectomy for acromegaly 6 years before admission. The erythrocyte sedimentation rate was 75 mm/hr, and the WBC count was 17,500/μl. Routine laboratory tests showed no abnormalities, and a test for antibodies to HIV was negative. Contrast-enhanced CT of the head showed a ringlike, enhancing, low-density mass in the right temporal lobe and a lytic defect in the right temporal calvaria (Figs. 1A and 1B). Minimal mucosal thickening in the right maxillary sinus was also present. MR of the head the next day confirmed the presence of an intraaxial mass in the right temporal lobe with mild midline shift. The mass was poorly defined and slightly hypointense to gray matter on T1-weighted images, 500/17/2 (TR/TE/excitations). Extension of the calvarial destructive lesion into the temporalis muscle was observed (Fig. 1C). T2-weighted images, 2500/90, showed a complex, mostly hyperintense 3-cm mass with an eccentric hypointense ring (Fig. 1D).

At surgery, a fleshy mass involving the temporalis muscle was found that extended through a small defect in the calvaria and dura. The mass directly invaded the temporal lobe. Resection of the temporal lobe component was attempted. Cultures of specimens from the mass were negative for mycobacteria, bacteria, viruses, and fungi. No microorganisms were seen on light or electron microscopic examination of histologic specimens. No further therapy was offered. The patient had an uncomplicated postoperative course, and follow-up examination 2 years later showed no recurrence of the mass.

Pathologic examination of the mass showed (1) a mixed cellular infiltrate containing neutrophils, eosinophils, and mono- and multinuclear histiocytes with lipid-laden cytoplasm and (2) erythrophagocytosis. Immunoperoxidase stains were moderately positive for S100 and strongly positive for T6, antigens used in the identification of Langerhans histiocytes. Despite the inability to identify Burbeck granules, the final diagnosis was histiocytosis X (eosinophilic granuloma).

Discussion

Histiocytosis of bone is well documented and can involve the calvarium. Histiocytosis X of the CNS most frequently involves the hypothalamus and can cause the classic clinical triad of diabetes insipitus, exophthalmos, and lytic skull lesions [2]. Solitary intracranial histiocytosis not involving the hypothalamus is rare, although at least six cases have been reported [3]. McCaffrey et al. [1] described 22 cases of histiocytosis X involving the middle ear and temporal bone. In our literature search, we found no report of any case with simultaneous involvement of brain, dura, calvaria, and extracranial soft tissues. Woodruff et al. [5] reported a constellation of findings similar to those of our case in a patient with glioblastoma multiforme. The initial possible causes in their case included sarcoma, meningioma, and metastasis. In our case, infection was a primary consideration. We suggest that histiocytosis X be included in the overall differential diagnosis for CNS lesions that have coexistent intra- and extracranial involvement.

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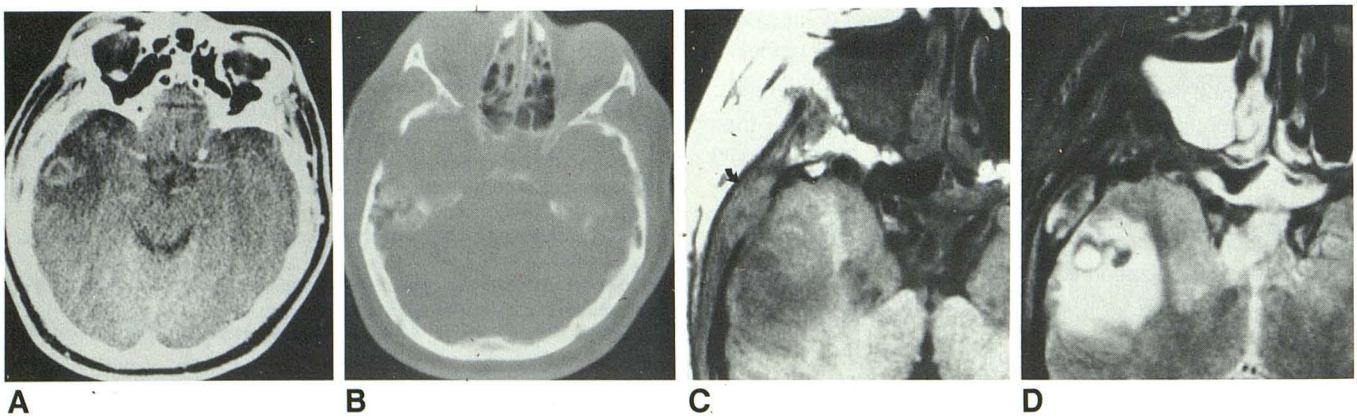

Fig. 1.—Histiocytosis X.
***A* and *B*, Enhanced CT scans show ringlike enhancing lesion (*A*) in right temporal lobe and, on bone window, right-sided calvarial destruction (*B*).**
***C*, T1-weighted MR image, 500/17/2, shows extracranial extension of lesion invading right temporalis muscle (*arrow*).**
***D*, T2-weighted MR image, 2500/90, shows extracranial extension and ringlike hypointense area in right temporal lobe corresponding to lesion in *A*.**