Solitary eosinophilic granuloma of the temporal lobe: case report and review of the literature

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Abstract A solitary eosinophilic granuloma of the central nervous system is an unusual manifestation of histiocytosis X. A unique case of a solitary eosinophilic granuloma of the right temporal lobe without osseous involvement is described. A 20-year-old man presented with a grand mal seizure. Magnetic resonance imaging demonstrated an intracranial enhancing mass in the right temporal lobe with marked vasogenic edema. A right temporal craniotomy was performed for resection of the lesion and the diagnosis of an eosinophilic granuloma was confirmed by histopathology. Follow-up MR imaging obtained 5 years following resection demonstrated no recurrence. Solitary eosinophilic granuloma should be considered in the differential diagnosis of enhancing mass lesions affecting the central nervous system. Although the natural history of solitary eosinophilic granulomas remains poorly defined, surgical treatment still remains the mainstay of therapy for these unifocal cerebral lesions.

Key words Brain · Eosinophilic granuloma · Histiocytosis · Magnetic resonance imaging (MRI) · Temporal lobe

Introduction

In 1953, a classification system grouped eosinophilic granuloma, Hand–Schüller–Christian disease, and Letterer–Siwe disease as one disease entity, histiocytosis X. This multisystem disorder of unknown etiology is characterized by a vast clinical spectrum ranging from a self-limited osteolytic skull lesion to patients with a fulminating disseminated systemic process. A triad of lytic skull lesions, exophthalmos, and diabetes insipidus has been described in Hand–Schüller–Christian disease. However, unifocal brain involvement in the absence of systemic disease or lytic skull lesions is quite rare (see Table 1). We now report a case of a solitary eosinophilic granuloma of the right temporal lobe without osseous involvement, and discuss the clinical presentation, appearance on magnetic resonance imaging (MRI), histopathology, and therapy.

Clinical summary

A 20-year-old right-handed male presented following a grand mal seizure. This event was preceded by a month-long history of episodic olfactory and gustatory sensations. They occurred several times a day, and were always followed by a bitemporal headache. He was seen by his family physician after the symptoms progressed to include sporadic nausea and vomiting as well as progressive right-sided headaches. A diagnosis of a viral syndrome was made, and the patient was discharged. On the morning of admission to the emergency room, the patient suffered a witnessed generalized tonic–clonic seizure lasting approximately 3 min with spontaneous resolution. Past medical history was unremarkable except for dental surgery 2 months prior to admission. The patient denied a recent history of fevers, weight loss, or exotic travel. There were no risk factors for human immunodeficiency virus (HIV) infection. The physical and neurological examinations were normal. The patient was afibrile and a fundoscopic examination revealed no papilledema.
A computed tomography (CT) scan demonstrated a 3.5 cm × 2.3 cm area of low attenuation in the right temporal lobe associated with marked vasogenic edema as well as uncal and subfalcine herniation. There was no associated osseous involvement. Following intravenous infusion of ionic contrast, there was marked enhancement of the mass within the temporal lobe (Fig. 1A,B). An MR image confirmed a uniformly enhancing intraaxial mass in the right temporal lobe (Fig. 1C).

A right temporal craniotomy was performed under general anesthesia with the aid of a frameless stereotaxic navigational device. A cortical incision made through the middle temporal gyrus revealed a mass 3 cm below the surface which was well encapsulated and extremely fibrous. The brain surrounding the lesion was extremely edematous and the mass lesion was resected in its entirety. An attachment of the lesion to the ependyma of the temporal horn was noted mesially, but no obvious vascular pedicle was seen.

The patient’s postoperative course was uneventful. Follow-up MR imaging 5 years following resection showed no evidence of recurrence of the lesion or systemic disease.

Pathologic findings

The histopathology was consistent with an eosinophilic granuloma (Fig. 2A–D). On hematoxylin and cosin (H&E) stained slides there was an abundance of angiocentric lymphoid cells comprising predominantly T cells and histiocytes, but also including B cells, macrophages, and focal aggregates of eosinophils in a background of densely collagenous stroma. These inflammatory cells were positive by immunocytochemistry for leukocyte antigens CD2 (T cells), CD5 (T cells), CD45 (common leukocyte antigen; lymphocytes and macrophages), and CD43 (Leu22; T cells and macrophages), and uniformly positive for S100 protein, but were glial-fibrillary acidic protein (GFAP) negative. Multinucleated giant cells were present throughout the specimen, but there was no necrosis. Flow cytometry confirmed a low lymphoid proliferative rate, and gene rearrangement studies did not reveal evidence for B- or T-cell clonal populations. The gram stain on the intraoperative specimen showed occasional white blood cells and no organisms. Stains and cultures for bacteria, acid-fast bacilli, and fungi were all negative. Fluorescent antibody studies for viruses (e.g. Cytomegalovirus, Varicella zoster, Herpes simplex) and toxoplasmosis were also negative. Electron microscopy was attempted on paraaffin-embedded, formalin-fixed tissues, which confirmed a mixed infiltrate of histiocytes, lymphocytes, and leukocytes in a collagenous and edematous stroma. No unequivocal Langerhans cell granules were identified.

Discussion

Histiocytosis X is a disease complex that includes Letterer-Siwe disease, Hand-Schüller-Christian disease, and eosinophilic granulomas. Solitary monostotic or polyostotic eosinophilic granulomas usually cause lytic osseous lesions, affecting most commonly the skull, femur, vertebrae, and pelvis. Histiocytosis X can affect reticuloendothelial cells anywhere in the body. Therefore, multiple lesions may involve the skin, liver, spleen, or even the gastrointestinal tract.15 Central nervous system (CNS) involvement of histiocytosis X, without concomitant osseous involvement, is extremely rare. However, isolated reports of involvement of the cranial vault, optic chiasms, orbits, hypothalamus (Gagal's granuloma), or pituitary do exist.1 Extrahypo-