Primary Eosinophilic Granuloma of the Optic Chiasm: Case Report

Optik Kiazmanın Primer Eozinofilik Granülomu: Olgu Sunumu

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Abstract: It is extremely rare to see eosinophilic granuloma of the central nervous system without osseous or multisystemic involvement. Only two adult cases with isolated cranial nerve involvement have been reported to date. This report describes an adult patient with isolated eosinophilic granuloma of the optic chiasm. We suggest that eosinophilic granuloma should be included in the differential diagnosis of tumors involving the cranial nerves, and particularly the optic chiasm.

Key words: Eosinophilic granuloma, histiocytosis X, cranial

INTRODUCTION

Eosinophilic granuloma (EG) is a disorder characterized by chronic non-neoplastic granulomas that contain proliferating histiocytes, plasma cells, and eosinophilic inflammatory cells. It is considered to be a unifocal or multifocal form of the diseases associated with Langerhans’ cell histiocytosis. The etiology of EG remains uncertain, but some believe that it involves an undefined immunologic disturbance caused by an unknown antigen (6). The lesions may be found in the bone marrow, skin, oral mucosa, retro-orbital tissue, lymph nodes, spleen, liver, lung, gastrointestinal tract, and central nervous system (CNS) (13).

In EG involving the CNS, the three most common sites of occurrence are the cranial vault, the suprasellar region, and the spinal column, respectively (1). It is extremely rare to find EG in the CNS without any osseous or multisystemic involvement, and only two cases of isolated cranial nerve involvement have been published to date (13,24). In this report, we describe an unusual case of primary EG of the optic chiasm, and stress the need to include this condition in the differential diagnosis for lesions affecting the cranial nerves.
A 25-year-old woman presented with a history of bilateral blurred vision of 4 months’ duration. She also reported having exaggerated thirst in the previous 2 months, with the signs of polydipsia and polyuria. There was no history of galactorrhea, amenorrhea, weight gain, sleep pattern disturbance, or changes in affective behavior or memory. Her physical examination on admission was normal, but a neurological examination demonstrated marked bilateral optic atrophy, severely reduced visual acuity in both eyes (0.1), and bitemporal hemianopsia. The results of routine laboratory studies were normal, apart from an elevated erythrocyte sedimentation rate (43 mm/h) and eosinophilia (7%). She was also diagnosed with diabetes insipidus of central origin.

Radiological Examination
Cranial computerized tomography (CT) revealed a 2x1.5 cm mass lesion in the suprasellar cistern. The lesion was isodense on non-contrast images, and showed marked contrast enhancement. Magnetic resonance imaging (MRI) revealed that the lesion involved the optic chiasm. It was isointense on T1-weighted images and hyperintense on T2-weighted images. We also noted marked homogeneous enhancement after the administration of gadolinium diethylenetriamine penta-acetic acid (Gd-DTPA). MRI also showed that the lesion extended to the pre-chiasmatic and post-chiasmatic portions of the optic tract. The cranial portion of the infundibulum could not be distinguished from the optic chiasm or from the mass lesion itself, whereas the caudal portion was normal in size and shape, and showed a normal contrast enhancement pattern (Figure 1). Aneurysm was ruled out by magnetic resonance angiography.

Surgical Procedure
The lesion was approached via a right pterional craniotomy. We opened the sylvian fissure and gently retracted the frontal lobe. On inspection, the optic chiasm was enlarged and appeared grayish brown in color. We dissected the optic chiasm free from the adjacent tissues, and made a midline incision in the optic chiasm parallel to the nerve fascicles. The lesion was firm, 2x1.5 cm, moderately vascularized mass that was invading the nerve tissue. There was no grossly apparent hypothalamic involvement. Operative dissection showed that the tumor was interfering with the continuity of almost all the nerve fascicles in the chiasm. As a consequence, only subtotal resection was possible.

Pathological Examination
The resected specimen was so small that we were unable to divide it for light and electron microscopic studies. Histological examination revealed a mixed inflammatory infiltrate including lymphocytes, plasma cells, eosinophils, and histiocytes. The infiltrating cells tended to be denser near blood vessels. High-power magnification showed that some of the histiocytes had grooved nuclei. Immunohistochemical studies revealed that the histiocytes were positive for S-100, but negative for glial fibrillary acetic protein (GFAP). On this basis, we labeled them Langerhans’ cells (Figure 1).

Treatment and Follow-up
Postoperative neurological examination revealed deficits identical to those found preoperatively. After the histopathological diagnosis was established, the patient was assessed for signs of skeletal and systemic involvement. She underwent hematological analysis, abdominal ultrasonography, CT, a skeletal radiographic survey, and radionuclide...
bone scanning, but no abnormalities were found. Low-dose radiation therapy (20 Gy in 10 fractions over 2 weeks) was administered, after which the patient was discharged. Follow-up MRI at 2 months post-surgery showed no obvious decrease in the size of the mass. The patient died at 31 months after surgery due to brain invasion by the tumor.

**DISCUSSION**

Eosinophilic granuloma is part of the spectrum of diseases associated with Langerhans’ cell histiocytosis. Histiocytosis X is the term that was first coined by Lichtenstein in 1953 (8) to describe a heterogeneous group of disorders. Today, this group is considered to include Hand-Schüller-Christian disease, Letterer-Siwe disease, and EG of the bone (7).

Gagel first described involvement of the CNS in histiocytosis X in 1941 (9). At that time, only a small number of cases of primary EG in the CNS had been reported. More recently, investigators have published larger series of patients with EG in the CNS, but these cases have also featured osseous or multisystemic involvement (11,12,17). The hypothalamus is often affected in cases of disseminated histiocytosis X, but isolated histiocytosis X of the hypothalamus is very rare (2,21,27). Infiltration of the hypothalamus or pituitary gland or both has been reported in 50% of histiocytosis X patients at autopsy (5,11,14,28). Diabetes insipidus is an important initial symptom when this structure is involved (14). Similar to the case reported by Smolik et al (24), in our patient the hypothalamus did not appear to be involved on radiological examination or at surgery.

Our patient’s preoperative CT scanning showed an isodense lesion with marked contrast enhancement. There was no surrounding edema. These findings were similar to those noted in the case of EG of the oculomotor nerve reported by Hardenack et al. (13). In previously described cases of solitary cerebral eosinophilic granuloma, the lesions have appeared either hypodense or isodense on CT (4,11,20,23). In our case, MRI revealed isointensity on T1-weighted images and hyperintensity on T2-weighted images, and this was in line with other reported findings. However, we observed marked homogeneous enhancement of the lesion after Gd-DTPA administration. This contrasts with other authors’ results (10,22), but confirms other more recent observations (6,13,15). Initially, we suspected that the mass was a glioma based on its location, signal, and contrast enhancement pattern. Our findings indicate that the radiological features of intracranial EG are highly variable, and we recommend that diagnosis of this type of lesion should not be based on radiological studies.

Since all the extirpated tissue was fixed in formalin, it was not possible to perform ultrastructural studies and identify Langerhans’ cells. Had it been possible to do electron microscopy, we might have been able to demonstrate Birbeck’s granules. However, the characteristic histological and immunohistochemical findings were sufficient to establish the diagnosis. These findings included the presence of many eosinophils, grooves in the histiocyte nuclei, cytoplasmic S-100 positivity, and GFAP negativity that excluded the possibility of a glial origin.

Treatment for EG remains controversial. Some authors suggest observation only (19). In the literature, radio- or chemotherapy is generally not recommended, provided that the lesions are localized, and are totally removed surgically (6). However, some researchers do advocate radiotherapy after total excision (12). Although EG is moderately radiosensitive (21), the optimal dose of radiation is still under debate. For multifocal EG, one report has stated that neither radiation nor steroid therapy is effective (8). Still, most cases involving subtotal resection and biopsy, or multifocal disease have been treated with radiotherapy (1,21,27), radiotherapy plus chemotherapy (7) or chemotherapy alone (25,16). The effects of chemotherapy and the optimal combination of chemotherapeutic agents are also unclear (7). Smolik et al. (24) administered a midline dose of radiation therapy (2700 r over 3 weeks) to their patient’s optic chiasm. Follow-up examination at approximately 8 months post-surgery showed that this was effective. Low-dose radiation has also been reported to cause regression of hypothalamic EG (26). However, we observed no positive effect of low-dose radiation therapy at the early postoperative stages in our patient. She died at 31 months after surgery due to brain invasion by the tumor.

The case we have presented contributes little new information to what is known about radiological diagnosis, histological diagnosis, and therapeutic approaches for EG involving the CNS or specifically the optic chiasm. However, it does underline the need to include EG in the differential diagnosis for cranial nerve lesions, particularly those involving the optic chiasm.
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REFERENCES


The common solitary lesion of the skull is known as eosinophilic granuloma, and the multiple lesions of the skull that are often associated diabetes insipidus are part of Hand-Schüller-Christian Syndrome.