Focal histiocytosis X of the parietal lobe

Case report

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The authors describe a case of histiocytosis X of the parietal lobe presenting as a space-occupying lesion on a computerized tomography scan of the brain. The clinical, radiographic, and therapeutic aspects of the case are discussed. A brief review of the literature is presented.

KEY WORDS — histiocytosis X • eosinophilic granuloma • Letterer-Siwe syndrome • Hand-Schüller-Christian syndrome • parietal lobe • radiation therapy

Histiocytosis X is a relatively rare disorder of the reticuloendothelial system involving the proliferation of histiocytes, granulation tissue, and inflammatory cells in many different organ systems of the body. Involvement of the central nervous system is rare, and is usually localized in the hypothalamus and occasionally in the pituitary gland (Gaget's granuloma). Disease affecting the central nervous system other than the hypothalamus is infrequently reported in cases of multifocal eosinophilic granuloma, but these have been autopsy studies. In this case of histiocytosis X of the parietal lobe, the lesion was demonstrated preoperatively by computerized tomography (CT) and angiography.

The surgical and pathological findings will be presented, with a brief review of the literature.

Case Report

This child was first noted in October, 1975, at the age of 26 months, to have a nonpulsatile mass on the left side of her head. A lytic lesion in the left temporo-parietal area was demonstrated on a skull film. The lesion was excised and the diagnosis of histiocytosis X was established on the basis of histopathological findings. An electroencephalogram at that time was normal. The disease was treated with local irradiation, cyclophosphamide, and prednisone. Subsequently, the disease progressed to involve multiple bones, namely the right orbit, right sixth rib, left seventh rib, right iliac crest, left humerus, and the T-10 vertebra. The lesions in T-10 and the left humerus were treated with irradiation (cobalt-60 source), delivering a total tumor dose of 800 rads and 1000 rads to the respective areas. Additionally, chemotherapy courses of vinblastine and prednisone were administered as described by Lahey.

Admission. In December, 1977, the patient presented with left-sided Jacksonian seizures, left homonymous hemianopsia, and papilledema. The skull film showed a lytic lesion in the right parietal bone and separation of the sutures secondary to increased intracranial pressure. A CT scan of the brain (Fig. 1) showed a large enhanceable mass in the right parietal lobe surrounded by marked edema, and also a shift of the midline structures to the left. Additional bone defects were seen in the clivus and sphenoid wing on the CT scan only. Cerebral angiography revealed an avascular mass in the right parietal lobe.

Operation. A parietofrontal craniotomy was performed. The location of the mass corresponded to the area of contrast enhancement on the CT scan. The borders of the tumor were poorly defined, and it seemed to infiltrate into the adjacent parenchyma. The tumor extended very deep. There was edema of the surrounding brain. A frozen section biopsy was compatible with histiocytosis X. A subtotal excision of
the tumor was carried out. The meninges between the mass and the skull bones were found to be free of the disease.

Pathological Examination. The excised tumor mass measured 4.0 \times 2.8 \times 2.2 \text{ cm}. The surface was irregular and varied from grayish-tan to brownish in color. The mass was firm in consistency. On sectioning, it was solid with no evidence of necrosis or hemorrhage. The cut surface was grayish-tan, with focal areas of brownish discoloration. Histologically, the cerebral tissue was infiltrated by neoplastic histiocytes, fibroblasts, atypical lymphocytes, and occasional eosinophils. The histiocytes were moderately large cells with a very well defined cytoplasmic border, and one or more rounded or indented vesicular nuclei.

There was evidence of collagen and reticular deposition in the tumor. The histiocytic infiltrate was similar to that seen in the right parietal bone lesion.

Postoperative Course. The postoperative CT scan of the brain showed evidence of residual tumor. The patient was treated with radiation (1600 rads with a cobalt-60 unit to the entire brain), dexamethasone and vinblastine. However, because of the lack of response shown by persistence of the lesion in the follow-up CT scan of the brain, an additional 1600 rads of irradiation was delivered to the entire brain. Follow-up CT scans of the brain showed a gradual decrease in the size of the residual tumor, eventually leading to complete disappearance (Fig. 2). The patient has remained neurologically well at 20 months posttherapy.

Discussion

Histiocytosis X has been described as a "non-neoplastic disorder of unknown etiology, pathogenically akin to an inflammatory reaction with multiple clinical manifestations mirroring the widespread distribution of the histiocytic system in the body." The common term "histiocytosis X" generally applies to clinical syndromes of eosinophilic granuloma, Hand-Schüller-Christian disease, and Letterer-Siwe disease; however, there have been dissenting opinions against this unified concept. Histiocytosis X is most commonly seen in children. Of the 73 patients with histiocytosis X treated at the Mayo Clinic over a 55-year period, only 15 were over 15 years of age.

Neurological complications of the disease are rare. In its classical form, it involves the hypothalamic-pituitary axis, causing exophthalmos, diabetes insipidus, visual loss, and other neurological disturbances. Focal involvement of the nervous system other than the hypothalamus is only rarely reported, and all these cases have been autopsy reports. Intracranial involvement is usually due to the extension...
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of the disease from the adjacent meninges. Primary intraparenchymal origin is a rare occurrence and is believed to be due to proliferation of the reticuloendothelial cells of the brain. A solitary lesion of histiocytosis X involving the cerebral cortex has been reported by Sivalingam, et al., but their patient underwent three consecutive craniotomies for apparent infarction of the brain before diagnosis was established.

The present case highlights the following observations:

1. The dominant findings on the CT scan are an intensely enhanceable lesion with extreme edema spread for a considerable distance in a finger-like fashion. The effect of therapy can be evaluated by follow-up CT scans of the brain.

2. The tumor is poorly demarcated, and a wide excision margin is required.

3. As opposed to the observation of marked radiosensitivity of the disease elsewhere in the body, some radioresistance may be encountered when it is located in the cerebral cortex. In the present case, in contrast to the observation of Sivalingam, et al., while the lesion in bone responded to irradiation at 800 to 1000 rads, a total of 3200 rads had to be delivered to the entire brain before the CT scan showed decrease in the size and eventual disappearance of the residual disease (Fig. 2).

References


