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Presumed Choroidal Langerhans Cell Histiocytosis Following a Previously Resected Solitary Central Nervous System Lesion in an Adult

Langerhans cell histiocytosis (LCH) is a rare disease of the mesenchymal dendritic cell system caused by a clonal proliferation of dendritic cells with Langerhans cell characteristics that may affect any organ, often with multisystem involvement and predominantly in children. Langerhans cell histiocytosis exhibits the characteristics of chronic inflammation, with typical granulomas consisting of CD1a-positive Langerhans cells, macrophages, T lymphocytes, and multinucleated giant cells and eosinophils. The most common central nervous system manifestation in LCH is involvement of the hypothalamic-pituitary region. Cerebral parenchymal involvement outside of the hypothalamic-pituitary axis by LCH is rare and may occur in the setting of multifocal systemic disease. Solitary unifocal LCH involving cerebral parenchyma is exceptionally rare. Ocular involvement in LCH is well recognized, but it normally manifests as eyelid or orbital infiltration and may involve multiple ophthalmic sites as part of a multisystem disseminated disorder.

We report a case of an adult previously diagnosed with a solitary left frontal LCH lesion who developed a solitary choroidal mass in his left eye believed to represent another LCH lesion 4 years later. We describe the clinical features of the lesion and the rapid response to external beam radiotherapy with resolution of visual symptoms.

Report of a Case. A 29-year-old man came to the emergency department with a 4-day history of sudden onset of metamorphopsia in his left eye and a reduction in subjective visual acuity. His right eye was asymptomatic. He had no previous ophthalmic history of note and was emetotropic. Four years previously, he had a grand mal seizure. Neuroimaging detected a solitary 1.5-cm lesion at the gray matter–white matter interface of the posteromedial region of the left frontal lobe that had a pattern of ring enhancement following gadolinium administration. No other lesions were identified. Complete excision had been achieved via a left frontal craniotomy. Histopathologic analysis revealed a polymorphous cellular infiltrate with marked perivascular sclerosis. Immunocytochemistry revealed that the cellular population was CD1a and S100 protein positive. Ultrastructural evaluation confirmed a population of cells with irregular, notched, and eccentrically placed nuclei, relatively abundant cytoplasm, and well-formed endoplasmic reticulum. A small number of Birbeck granules were identified in this population of cells, and a diagnosis of solitary cerebral LCH was made. Throughout the following 4-year period, repeat magnetic resonance im-

Figure 1. Magnetic resonance image of the coronal section of the head. A single Langerhans cell histiocytosis lesion that is enhanced after gadolinium administration appears in the posteromedial region of the left frontal lobe.
aging scans of the head showed no evidence of disease recurrence, and the patient had been well ever since.

On ophthalmic examination, visual acuity was 6/12 OS and 6/6 OD. Both anterior segments appeared normal and had normal intraocular pressures. Examination of the left fundus revealed a solitary, raised choroidal lesion measuring approximately 5 disc diameters in area (Figure 4). The lesion was elevated, with a creamy yellow appearance overlying the superotemporal vascular arcade, and causing distortion of the fovea. B-scan ultrasonography showed the raised lesion (raised approximately 2 mm) with low-medium reflectivity. Fluorescein angiography revealed early masking of the underlying choroid with late leakage overlying the lesion. Optical coherence tomography confirmed subretinal exudation. Repeat neuroimaging of the head revealed no evidence of regrowth of the tumor from its original site. In addition, repeat medical examination and skeletal survey revealed no other systemic focus of disease activity. In view of the patient’s history, a diagnosis of a solitary choroidal LCH lesion was made. Low-dose (1000 rad [10-Gy]) external fractionated radiotherapy to the lesion was performed. Within 2 weeks of treatment, there was a rapid improvement in both visual symptoms (visual acuity, 6/6 OS) and clinical appearance (Figure 5). The patient is still being followed up.

Comment. To our knowledge, this is the first reported case with a choroidal LCH lesion developing after a previously resected and completely excised solitary cerebral parenchymal lesion. The original diagnosis was based on the result of the excisional biopsy of the solitary cerebral lesion demonstrating a proliferation of histiocytic cells of Langerhans type in addition to the expression of CD1a, S100 protein, and Birbeck granules on electron microscopy.11 In non-LCH, CD1a is only very rarely positive. Positivity for S100 is commonly sought in the evaluation of potential LCH lesions but is not specific and can be observed in histiocytes other than Langerhans cells. Applying criteria proposed by the Histiocyte Society, a definitive diagnosis of LCH requires demonstration of either the expression of CD1a on the cell surface or the ultrastructural identification of Birbeck granules.
The diagnosis of the fundal lesion as an LCH lesion was based on the clinical and angiographic appearance of the fundal lesion in addition to the ultrasonographic findings and rapid response to external beam radiotherapy. We did not consider it practical or prudent to perform a biopsy owing to the risks involved and the location of the lesion.

Owing to the rarity of the condition, there is no definitive consensus as to the optimum treatment of solitary ocular LCH. Some LCH lesions can be observed and have been shown to spontaneously regress. However, owing to the sudden onset of symptoms and the location close to the fovea in our case, the LCH lesion was treated with an initial course of low-dose external beam radiotherapy and the response was observed. The lack of any other focus of disease precluded the need for systemic chemotherapy, and the lesion was not surgically resectable owing to its location.

In conclusion, to our knowledge, we report the first case of a solitary choroidal LCH lesion developing in an adult some years after the patient had a solitary cerebral parenchymal LCH lesion surgically resected. This case shows that many years after complete surgical excision of a solitary cerebral LCH lesion, secondary foci of disease can still occur. Although rare, LCH must be considered in the differential diagnosis of any choroidal mass.