We treated a 46-year-old Japanese man with Langerhans cell histiocytosis (LCH) localized to the eyelid alone. He was cured successfully by local and complete resection. Results of pathological examinations of the excised tumor demonstrated diffuse infiltration by atypical histiocytic cells with eosinophilic cytoplasm and convoluted nuclei, S100 immunoreactivity, and tennis-racket–shaped Birbeck granules. Based on these pathological findings, we diagnosed LCH. Clinical examination revealed no LCH involvement in other parts of the body. To our knowledge, there has been only one report of LCH occurring as an isolated tumor in the eyelid. Generally LCH has been reported in children or young people. This is an unusual case of LCH isolated to the eyelid of an older patient.

Langerhans cell histiocytosis (LCH), formerly referred to as histiocytosis X, is a tumorous, proliferative disorder of Langerhans cells. It frequently involves bone, spleen, lymph nodes, and skin. The lack of reliable blood serum or histopathological markers of LCH makes accurate prognosis difficult. Some patients get better without treatment, while others worsen rapidly and die. In general, good prognoses seem to be related to late onset and to the absence of lesions at multiple visceral sites. The eyelid is seldom involved; only a few such cases have been reported in the English- and Japanese-language literature, and in all of them, except one, other structures were affected as well. In the case report about histiocytosis X localized to the eyelid, the diagnosis was based on the results of histological and immunohistochemical examinations, but without the demonstration of the presence of Birbeck granules by electron microscopy. We report a case of LCH localized to the eyelid alone, definitively diagnosed by histological, immunohistochemical, and electron microscopic examinations.

REPORT OF A CASE

A 46-year-old man complained of left upper eyelid swelling with foreign body sensation and lacrimation lasting for 3 weeks. No history of trauma, chronic inflammation, or relevant family history was noted. Visual acuity was 20/15 OD (not correctable), and 20/30 OS (correctable to 20/15). A solid nodule measuring 7 mm in diameter was found beneath the eyelid skin, and a decubital ulcer was observed at the center of this lesion (Figure 1).

Results of a histological examination of an incisional biopsy specimen showed features consistent with LCH. General examination revealed no lymphadenopathy or hepatosplenomegaly, and no eruptions or nodules suggesting the presence of LCH anywhere else on the skin. Results of routine blood tests were normal. Neither x-ray films of the skull and lung nor a computed tomographic scan of the truncus and head revealed any remarkable features. Based on these findings, we diagnosed the tumor as a focal type of LCH. With the patient under general anesthesia, we excised the lesion and reconstructed the upper eyelid. As no tumor cells were observed at the edge of the excised specimen, no follow-up chemotherapy or radiation therapy was prescribed.
Results of the histological examination of the tumor revealed ulceration of the overlying epidermis. Atypical histiocytic cells with convoluted nuclei and some multinucleated cells invaded the deep muscle layer. Many lymphocytes and eosinophils were present (Figure 2, A-C). Most of the atypical histiocytic cells were immunohistchemically intensely reactive to anti–S100 protein, highly specific to Langerhans cells (Figure 2, D), and somewhat reactive to \( \alpha_1 \)-antichymotrypsin, lysozyme, and KP-1 (CD68), possible markers for histiocytes. Transmission electron microscopy revealed Birbeck granules, the characteristic feature of Langerhans cells (Figure 3), and definitively identified the tumor as LCH.

Twelve months after surgery, there has been no recurrence in the eyelid or other structures.

**COMMENT**

Langerhans cells belong to the mononuclear phagocyte system. Histopathological examination alone, revealing a diffuse infiltration by atypical eosinophilic cells with large cytoplasm and convoluted nuclei, is usually adequate for the diagnosis of LCH. Immunohistochemistry helps to identify these cells as Langerhans, but transmission electron microscopy can confirm the diagnosis by identifying characteristic Birbeck granules.
microscopy revealing Birbeck granules definitively identifies Langerhans cells.

Langerhans cells normally occur in body surface tissues such as epidermis, conjunctiva, corneal limbus, and epithelium of respiratory tract. They are especially numerous in the dermis of normal eyelid margin skin and in tarsal subconjunctival tissue of patients with allergic conjunctivitis. Because they are rare in healthy tarsal conjunctiva, we suspect that the tumor in this case may have originated in the epidermis of the eyelid margin and progressed to the tarsal conjunctiva over the eyelid margin. Classification according to The Writing Group of the Histiocyte Society\(^1\) would categorize this tumor as a pure cutaneous histiocytosis of class I. This case is unusual because of the age of the patient; LCH usually occurs at an early age. We found nothing in this patient's history or examination results that would explain why LCH developed when he was in his 40s.

Irradiation, carcinostatic drugs, and steroid therapy are commonly employed in cases with bone involvement and/or hepatosplenomegaly. Irradiation, though very effective, risks subsequent development of central nerve disorders and secondary malignant neoplasms. Etoposide, the most effective carcinostatic drug for LCH, may cause secondary cancers, including leukemia. Font\(^6\) reported good long-term results when patients with isolated LCH were treated by excision of the lesion alone. Our patient's prognosis is probably good because of his relatively old age for patients with LCH, and because his LCH was apparently limited to a single, localized tumor; however, careful, prolonged follow-up is required.

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REFERENCES


Notes From Our Ophthalmic Heritage

A look at the past . . .

The Choroid and Retina

The ophthalmoscope has effected such a revolution in the opinions formerly entertained as to the morbid conditions of the choroid and retina, that the terms at the head of this Chapter have already lost much of the meaning they would have suggested a few years ago; and many cases, which then were vaguely spoken of as “functional Amaurosis,” could now be proved to depend upon actual changes of structure.