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Dear Editor,

LANGERHANS CELL HISTIOCYTOSIS OF THE EYELID

We present an unusual case of Langerhans cell histiocytosis (LCH) masquerading as a non-resolving chalazion, which was successfully treated with excision biopsy and intralesional corticosteroid.

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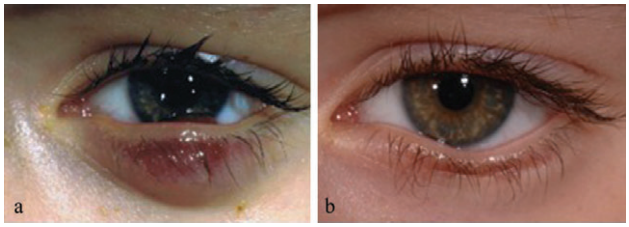

Fig. 1 (a) left lower lid lesion on presentation (b) left lower lid following excision and intra-lesional steroids.

A 7-year-old boy presented with a few weeks history of a left lower lid mass. This started as a tiny painless nodule and gradually increased in size. This was diagnosed as a chalazion and treated conservatively. As the lesion increased further in size, the patient was referred for further evaluation. There was no past medical history of any significant co-morbid conditions or localised trauma.

On examination, a dark red kidney bean-shaped mass measuring 1 cm × 0.75 cm was felt over the lower lid (Fig. 1a). The mass was non-mobile and extended up to the lid margin. There were no signs of infection or ulceration of the overlying skin. General examination revealed no lymphadenopathy or organomegaly. Initial haematological investigations were normal. Due to the unusual appearance of this lesion, an incisional biopsy was arranged.

Histopathological examination revealed the presence of chronic inflammatory infiltrate of large histiocytic cells with partly frothy eosinophilic cytoplasm, strongly positive for the marker s-100, consistent with the diagnosis of LCH. Computerised tomography (CT) scan of the head and its orbits revealed no orbital extension. Chest CT scan and skeletal survey were normal.

The staging process had revealed localised LCH affecting the left lower eyelid only. During the investigation period, the lesion increased in size to 1.5 cm in width, height and depth. Localised excision combined with intralesional injection of steroid was subsequently planned. A solid mass was found adherent to the anterior tarsal plate extending up to the lid margin. The tarsal plate was thickened. The lesion was debulked and 0.2 mL of 40 mg/mL triamcinolone acetonide was injected into the local tissues.

Upon review 2 days postoperatively, only minor irregularity of the lid margin was noted. There had been significant reduction in the size of the initial lesion. Further resolution occurred over the following two-week period (Fig. 1b). At three-month follow-up, there has been no evidence of recurrence.

Langerhans cells (LC) normally occur in body surface tissues such as epidermis, conjunctiva, corneal limbus, and epithelium of the respiratory tract. They are especially numerous in the dermis of normal eyelid margin skin where the tumour in this case would have originated.[1] LCH is characterised by an aberrant proliferation of LC and is part of a group of clinical syndromes known as the histiocytoses. The incidence of LCH is 4.6 per million children (age 0–14 years).[2]

LCH comprise a wide spectrum of clinical manifestations ranging from solitary lesions that usually exhibit benign clinical behaviour to widely disseminated lesions that may exhibit a malignant course. Tissues characteristically involved in LCH are bone, skin, lung, liver, spleen, bone marrow, lymph nodes and the hypothalamic-pituitary region, although involvement of other organs such as bowel can occur.[3]

Depending on tissue involvement, LCH has been traditionally divided into single-system and multisystem disease. In the case of single-system disease involving bone or lymphoid tissues, several sites may be affected, and the staging is then classified as multifocal, single-system disease. In the setting of multisystem disease, involvement of certain tissues – bone marrow, liver, spleen and lungs – so called risk organ positivity, are associated with a worse prognosis.[3]

The exact pathogenesis of LCH remains unknown. Debate exists as to whether this process is reactive or neoplastic.[4] Although clinical manifestations vary and often overlap, histopathological features tend to be similar. The hallmark of LCH is the presence of Birkbeck granules on electron microscopy.[5] The diagnosis of LCH is also confirmed by the presence of CD1a or CD207 (Langerin) positive histiocytic cells.[2]

In general, the treatment of LCH depends on the site and severity of the disease process. In the isolated skin lesions, minimal therapy with topical emollients may be sufficient. Carcinostatic agents, irradiation and steroid therapy are employed in cases with multisystem disease with bone involvement and/or haepatosplenomegaly.[4,5] Irradiation and chemotherapeutic agents, although very effective, risk subsequent development of secondary malignant neoplasms. In isolated LCH excision alone or in combination with topical oral steroid therapy, there have been found to induce complete remission, thereby avoiding irradiation and chemotherapeutic agents.

The use of intralesional corticosteroid in the treatment of Langerhans cell histiocytosis lesions has been previously advocated and especially used for osseous lesions.[3] In this case, we found the combination of surgical excision and intralesional steroid enough to induce complete remission, and therefore, avoiding long-term treatment with systemic steroids and their associated side effects.

In summary, LCH, although a rare disease entity, should be considered in the differential diagnosis of eyelid lesions. This case demonstrates that selected patients with unifocal cutaneous LCH may be a good candidate for a trial of intralesional corticosteroid therapy in combination with surgical excision. Careful long-term follow-up is required to ensure complete remission.

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