Langerhans cell histiocytosis in the frontal and zygomatic bones of an adolescent girl

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DESCRIPTION

An adolescent girl with no medical history presented to our institution with right upper eyelid swelling and pain. She initially went to an external ophthalmological clinic, where she was given eye medications due to the suspicion of ocular herpes. After receiving eye medications, her right eye swelling improved and relapsed. The girl had no history of fever or any other systemic illness. Clinical examination revealed right eyelid swelling and tenderness (figure 1A). Her visual acuity and field tests as well as ocular movements were normal. The results of all routine laboratory tests were within normal limits. Head CT revealed an osteolytic lesion in the right frontal and zygomatic bones (figure 2A–E). MRI revealed an exophytic, infiltrative, heterogeneously contrast-enhanced mass extending from the intraorbital space to the subcutaneous space (figure 2F–I). Due to the suspicion of Langerhans cell histiocytosis (LCH), other tumours of skull and abscess, biopsy was performed via a subeyebrow incision (figure 1B), and histopathological examination of the specimen was conducted. We found a soft, friable, haemorrhagic lesion within the eroded frontal bone (figure 1C). Microscopically, the lesion consisted of large histiocytes with abundant eosinophilic cytoplasm and oval, folded or grooved nuclei with interspersed eosinophils, lymphocytes and multinucleated giant cells (figure 1D). These typical histopathological features and immunochemical evidence of CD1a-positive (figure 1E) and S100-positive cells (not shown in the figure) confirmed the diagnosis of LCH. The postoperative course was uneventful, with no neurological deficits. Staging investigations, including whole-body CT and bone scans, were performed, but no evidence of distant disease involvement was found. The patient was subsequently referred to a paediatric oncologist. Based on these results, she was finally diagnosed with single-system LCH (SS-LCH) with multifocal bone (MFB) lesions. She is currently receiving chemotherapy with induction prednisolone, cytarabine and vincristine according to the LCH-19-MS/MFB protocol of the Japan LCH study group.[1] This treatment is set to last for 30 weeks. Complete recovery of her right upper eyelid swelling and pain occurred within 4 weeks.

LCH is an uncommon presentation in daily practice. Orbital LCH is extremely rare, accounting for approximately <1% of all orbital tumours.[2] The most common clinical manifestations of orbital LCH include unilateral orbital pain, headache or localised eyelid redness and swelling.[3] Due to the lack of understanding of the pathogenesis of LCH, optimal diagnosis and therapy remain challenging. Indeed, in this case, she was misdiagnosed with ocular herpes.

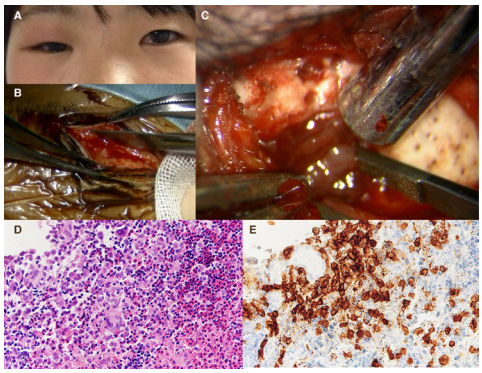

Figure 1 (A) Photograph on admission showing eyelid swelling of the right orbit. (B) Intraoperative photograph showing incision of skin biopsy. (C) Intraoperative photograph showing a soft, friable, haemorrhagic tissue within the eroded frontal bone. (D) Microscopic section of the tumour showing findings consistent with Langerhans cell histiocytosis. (E) Immunohistochemical staining showing strong positivity for CD1a.

The natural form of LCH varies, ranging from a slow, benign, localised bony or soft tissue lesion to a rapidly progressive, widespread, multisystemic fatal disorder. Patients with limited orbital involvement generally tend to have a good prognosis.[3 4] Minimal

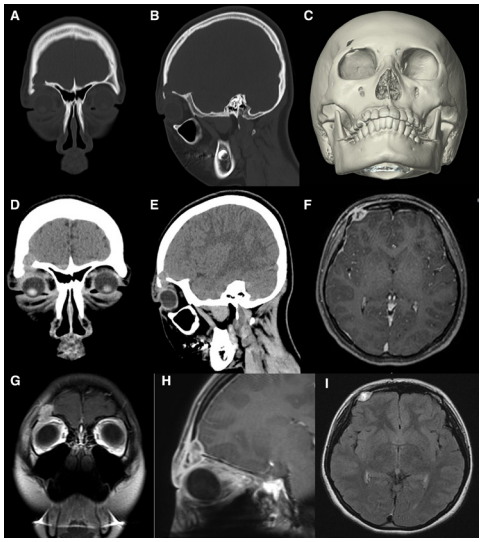

Figure 2 (A–C) Bone CT scans on admission showing an osteolytic lesion in the right frontal and zygomatic bones. (D,E) Head CT scans on admission. (F–H) Contrast-enhanced T1-weighted MRIs on admission showing a heterogeneously contrast-enhanced mass extending from the intraorbital space to the subcutaneous space. (I) Fluid-attenuated inversion recovery image on admission.

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intervention (surgical resection with or without intralesional corticosteroids) may be sufficient for patients with an isolated orbital LCH. However, the Histiocyte Society has suggested that the following disease extent categories be considered indications for systemic therapy[5]: SS-LCH with 'central nervous system-risk' lesions; SS-LCH with MFB; SS-LCH with 'special site' lesions; and multisystem LCH with/without the involvement of risk organs. Indeed, in this case, systemic chemotherapy was administered according to the final diagnosis of SS-LCH with MFB.

Patient's perspective

Father: 'We were informed that our daughter had to undergo a biopsy to establish a histopathological diagnosis of the orbital lesion. We agreed for the same. We were relieved with the improvement in symptoms. After discharge, she was fine and systemic chemotherapy was initiated.'

Learning points

- We describe an uncommon case of orbital Langerhans cell histiocytosis (LCH) in an adolescent girl who presented with right orbital pain and oedema of the upper right eyelid.
- If unilateral orbital pain and/or eyelid swelling is found in a patient with an orbital bone defect, then orbital LCH should be considered and included in the differential diagnosis.
- Suggested treatment for patients with orbital LCH varies according to the disease location.

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