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CASE REPORT

Case study: Langerhans' cell histiocytosis (LCH)

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Summary The clinical spectrum of Langerhans cell histiocytosis (LCH) can be very diverse. Pathology can range from a single bony lesion to systemic disease with multi-organ dysfunction. Up to 61% of patients with LCH have otologic involvement and in 5—25% it might be the initial form of presentation [A.J. Goldsmith, D. Myssiorek, E. Valderrama, M. Patel, Unifocal Langerhans' cell histiocytosis (eosinophilic granuloma) of the petrous apex, Arch. Otolaryngol. Head Neck Surg. 119 (1993) 113—116; R. Hermans, B. De Foer, M.H. Smet, J. Leysen, Eosinophilic granuloma of the head and neck: CT and MRI features in three cases, Pediatr. Radiol. 24 (1994) 33—36; T.V. McCaffrey, T.J. McDonald, Histiocytosis X of the ear and temporal bone: review of 22 cases, Laryngoscope 89 (1979) 1735—1742]. The otologic findings of LCH are similar to otomastoiditis and therefore LCH should be considered as a possible differential diagnosis in very young patients presenting with signs and symptoms suggestive of chronic otitis media.

1. Case study

A 2-year-old boy presented to our ENT clinic with a discharging right ear for 1 month. There was no history of previous ear infections or hearing loss. Apart from suffering from recurrent atopic eczema he had no significant medical history.

A complete ENT examination was performed. Examination of his right ear revealed a purulent discharge with a polyp in the deep part of the external ear canal, obscuring the tympanic membrane. He also had a small lymph node in the right postauricular area. Examination of his left ear was normal.

A diagnosis of otitis media was made and he was treated with oral Co-amoxiclav and aminoglycoside eardrops.

A week later, the ear canal polyp was still present and an inflamed fluctuant swelling was present in the postauricular area. The patient was apyrexial and he had no neurological deficits. A diagnosis of acute mastoiditis was made and a mastoidectomy was performed.

Intraoperative findings revealed an eroded bony external ear canal and mastoid bone giving the picture of an 'automastoidectomy'. The deep part

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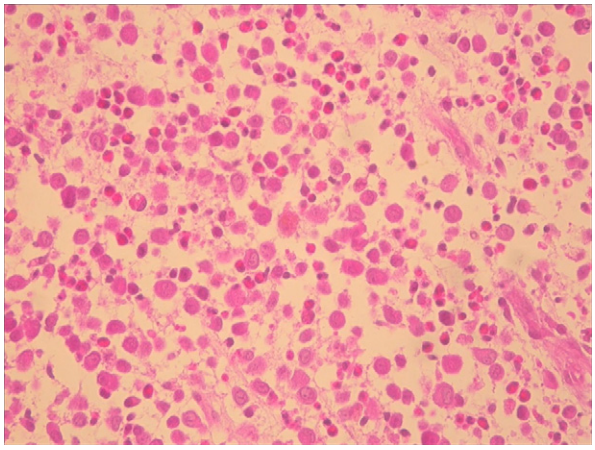

Fig. 1 High-power view demonstrating a proliferation of Langerhans cells. Scattered eosinophils are present in the background.

of the external ear canal, middle ear and mastoid cavity were filled with friable soft tissue and no tympanic membrane or ossicles could be identified. There was no evidence of cholesteatoma. Biopsies of the soft tissue were taken and sent for histology.

The histology result showed fragments of granulation tissue lined by focally ulcerated squamous epithelium. There was marked congestion of stromal blood vessels and the tissue was infiltrated by numerous eosinophils and histiocytic cells. Stains for fungi and bacteria were negative. The histiocytic cells stained positive for S100, CD68 and CD1a (Figs. 1—4).

A diagnosis of eosinophilic granuloma was made and the child was referred to the oncology service.

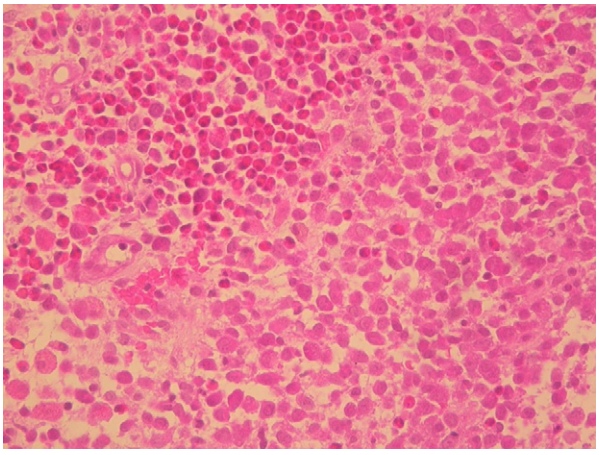

Fig. 2 Histiocytes, some of which are multinucleate, are also present in the background and these are highlighted well on the CD 68 stain (upper left-hand corner), high-power view.

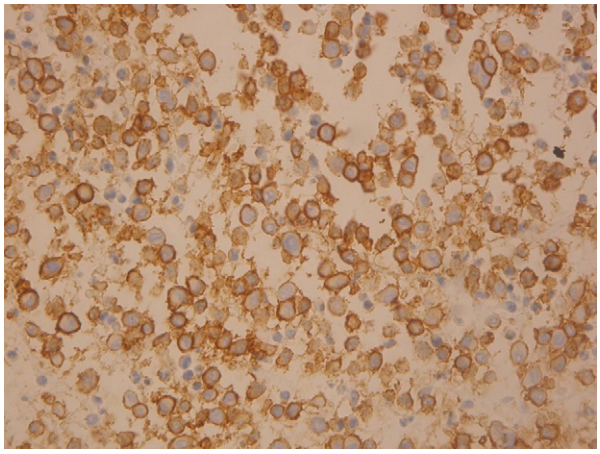

Fig. 3 The Langerhans cells show strongly positive with the CD1a stain, high-power view.

The patient was assessed by the oncologist. The child did not have polyuria or polydipsia (posterior pituitary involvement). Furthermore there was no history of anorexia, hyperphagia, changes in temperature control or sleep patterns (hypothalamic involvement) and he had normal anthropometry (anterior pituitary involvement). On examination he was well grown and had no focal neurology. This in-depth history is important because children with lytic lesions involving the temporal bone are at risk of developing CNS disease, in particular diabetes insipidus. The diabetes insipidus is classically unresponsive to systemic therapy and irreversible.

Complete systemic investigation including a temporal bone CT scan revealed this to be an isolated temporal bone lesion (Fig. 5). He was reviewed by the dermatologists to exclude cutaneous involvement by LCH. The skin disease was felt to be chronic atopic eczema rather than cutaneous LCH.

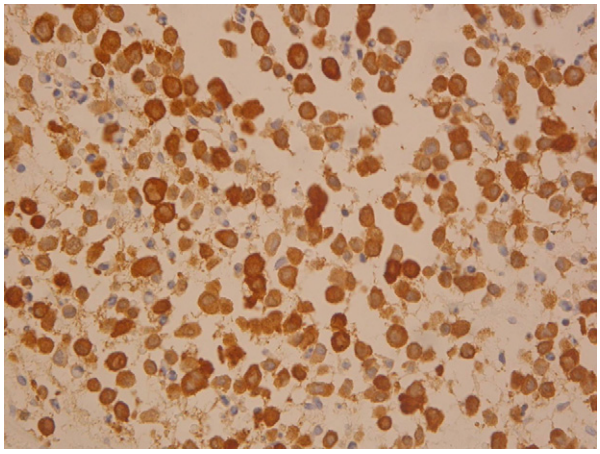

Fig. 4 Tumour cells are also positive for the s100 stain, high-power view.