Journal of Paediatrics and Child Health

25 April 2012

Dear Editor,

AN UNUSUAL TRAUMATIC ULCERATION OF THE TONGUE

We report a case of an 11-month-old girl who was referred to us for fever, difficult feeding with progressive weight loss and a painful ulcer on the ventral surface of the tongue. Her clinical history was characterised by a perinatal hypoxic-ischaemic brain injury. The intra-oral examination revealed an ulcerative lesions extending from the tip to the ventral surface of the tongue. It was ovoid, well circumscribed, with raised and indurated edges and with a yellowish fibrinous base (Fig. 1a,c – lower arrow). Biochemical analysis, immunological and genetic studies were normal; cultivation of the lesion revealed flora saprophyte. As neurological examination revealed a multifocal dystonia including abnormal movements of the oro-facial musculature that caused the impact of the tongue between the lower central incisors, the diagnosis of 'late Riga-Fede disease' (RFD) was considered. The electroencephalogram (Fig. 1b) showed bihemispheric slow frequency rhythms, while the brain magnetic resonance imaging showed a thinning of the

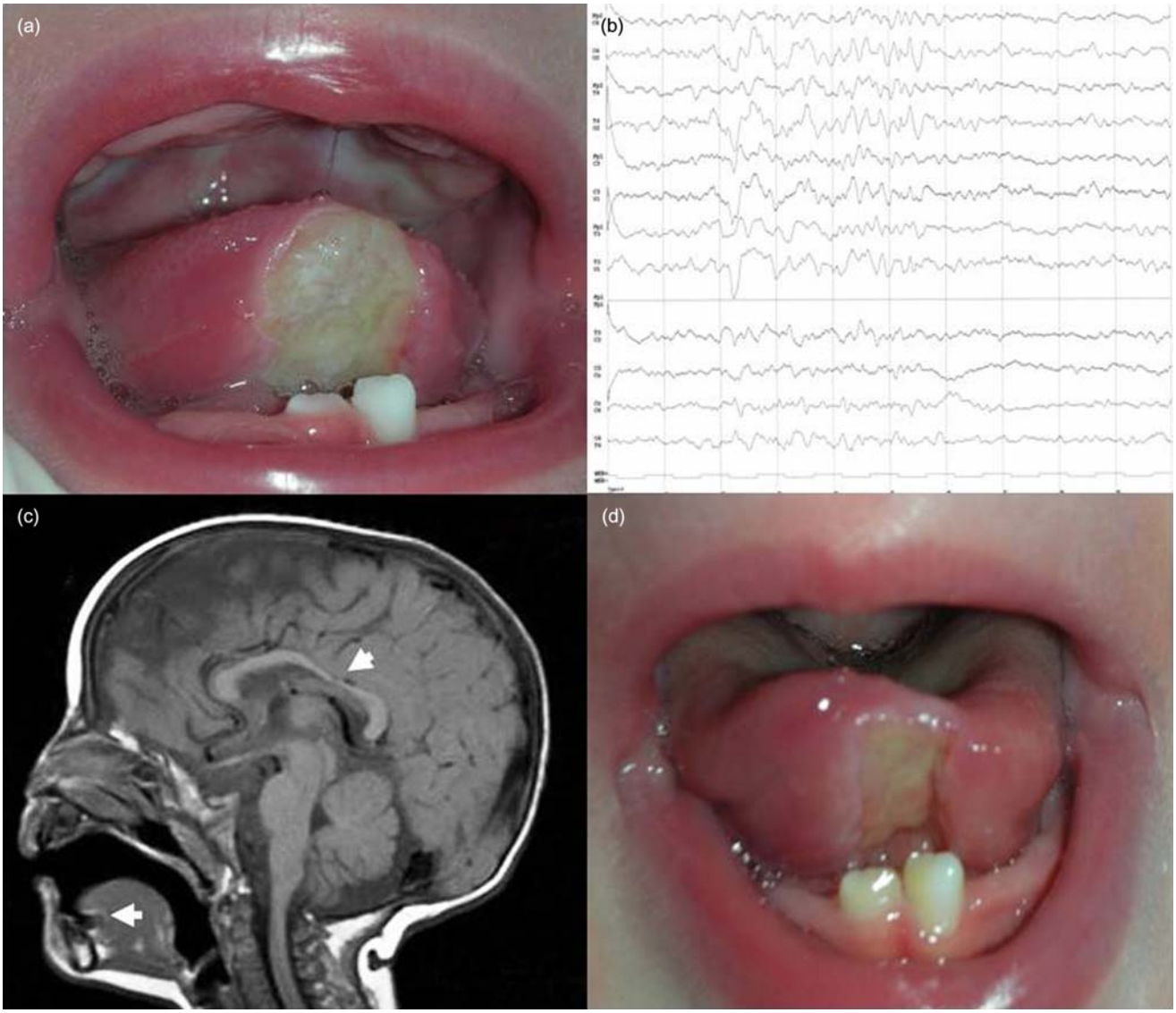

Fig. 1 (a) Clinical features of the oral ulcer: it appears ovoid in shape, well circumscribed, with raised and indurated edges and with a yellowish fibrinous base. (b) Electroencephalogram examination: it shows a bihemispheric slow frequency rhythms. (c) Magnetic resonance imaging showing a thinning of the *corpus callosum* (upper arrow) and the tongue ulceration (lower arrow). (d) Clinical amelioration of the lingual lesion after the treatment.

Conflict of interest: None declared.

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corpus callosum (Fig. 1c, upper arrow). Fever was treated with paracetamol, while the treatment with carbamazepine improved her neurological impairment, which when associated to a dental conservative approach (use of a flat nipple to avoid the impact of the teeth against the tongue associated to the topical application of a chlorhexidine 0.5% and hyluronic acid-based gels), led to a clinical amelioration of the ulcer (Fig. 1d) with an improvement of feeding and a gradual gain of weight.

RFD is a rare, oral, ulcerative condition described by Antonio Riga, an Italian physician in 1881. The oral ulcer is characterized by continuous traumatic injuries produced by natal or neonatal teeth, usually lower incisors, resulting in tongue ulceration.[1] This form is called 'precocious or early RFD', and appears before 6 months of age, a period in which the child is edentulous. However, a 'late RFD' appearing in 6 months of age or in older children is also described. It is not due to a precocious teeth eruption, but, as in our case, to neurological or developmental disorders in a child with normal chronology of dental eruption.[2,3] Hence, this latter disorder may be the initial presentation of some serious underlying medical problems, as Lesch–Nyhan syndrome, familial dysautonomia and/or other neurological disorders as parasomnia, hereditary chin trembling and facio-mandibular myoclonus that may cause nocturnal tongue biting.

From a point of view of its clinical manifestation, the differential diagnosis of RFD includes granular cell tumour, myofibroma, eosinophilic granuloma and other traumatic ulcers. Malignancies (sarcoma and lymphoma) and infectious diseases (acquired/congenital syphilis, tuberculosis and ulcerative fungal infections) must also be ruled out.

Because the precocious extractions may have a negative impact on the dentoalveolar growth of the children, the treatment of choice of RFD should be conservative (as in our case), reserving the surgical approach, with the extraction of the teeth and subsequent application of a space maintainer, only in case of non-healing in conservative treatment.

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10 November 2011

Dear Editor,

LANGERHANS CELL HISTIOCYTOSIS – A MIMICKER OF TUBERCULOSIS OF THE SPINE

Langerhans cell histiocytosis (LCH) and tuberculosis are mimickers in the bone.[1–3] We present an interesting case of an 8-year-old Indonesian-Chinese boy, who presented with 1 month of low back pain and thoracolumbar tenderness. Imaging studies demonstrated T10 vertebral plana with preserved disc spaces and a paraspinal soft tissue mass causing cord compression. Bone scan revealed increased uptake over T10 but skeletal survey was unremarkable. Tumour resection, posterior instrumentation and fusion of the spine were performed. Histology was consistent with eosinophilic granuloma.

He re-presented 3 years later with right neck/shoulder pain, right arm weakness and limited abduction for 8 months with no other systemic symptoms. With a positive household contact of pulmonary tuberculosis (pTB) in Indonesia 2 months prior, positive Mantoux test (18 mm) and imaging studies (Fig. 1), he was diagnosed with cervical spine tuberculosis. Blood counts, erythrocyte sedimentation rate and chest X-ray were unremarkable. He was treated with anti-tuberculosis therapy (rifampicin, isoniazid, ethambutol and pyrazinamide for 6 weeks, then isoniazid monotherapy) with no adverse side effects. He underwent laminectomy/drainage of cervical spine abscess within the first week of anti-tuberculosis therapy.

Intra-operatively, soft friable tumour tissue eroding C3 lamina, compressing the spinal cord, could not be resected completely. Histology confirmed fibrosis with reparative bone formation and focal areas of LCH, positively staining S100 and CD1A. Ziehl–Neelsen stain and tuberculosis cultures were negative. The patient was commenced on LCH-III protocol chemotherapy for multifocal bone disease. A re-evaluation magnetic resonance imaging spine at 6 months showed no disease progression.

This is an interesting case where a LCH spine recurrence was misdiagnosed as tuberculosis of spine due to a positive contact of pTB, positive Mantoux test and imaging studies. To highlight the pitfalls in diagnosis, in a patient from Indonesia where pTB is rampant, the likelihood of 'history of exposure' to tuberculosis in the community is very high and does not necessary equate to true tuberculosis exposure. Furthermore, the median time for reactivation of unifocal bone disease in LCH has been reported to be 1 year;[4] in this case, a second involvement of another vertebra after several years is uncommon, which might have been an added challenge.

We highlight some indicators which may help in distinguishing the two conditions:

1 *Radiological investigations.* Presence of vertebral plana with preserved disc spaces is typical of LCH, while the disc spaces will be lost in tuberculosis of the spine.[5] Despite the presence of vertebral plana with preserved disc spaces, the history of pTB contact and positive Mantoux test led to the misdiagnosis of tuberculosis spine in this patient.

Conflict of interest: None.