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Congenital facial nerve agenesis

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Abstract

We present a case of a seven-year-old child with a congenital facial palsy, diagnosed at birth, who subsequently developed a non-tuberculous mycobacterial (NTM) infection of the ipsilateral parotid gland. This required parotid exploration to treat the NTM disease with the intention of identifying and protecting the facial nerve to preserve any residual facial nerve function. At operation, thorough exploration revealed the complete absence of the nerve both at the stylomastoid foramen and more peripherally within the substance of the parotid gland. Exploration of the facial nerve for congenital facial paralysis is not normally indicated. Surgical treatment, if required, tends to involve the use of techniques such as cross facial nerve and free vascularized muscle grafting. To our knowledge this is the first reported case of complete congenital facial nerve agenesis, diagnosed incidentally during a surgical procedure for an unrelated condition.

Key words: Facial Paralysis, Congenital; Mycobacterium, Non-tubercular; Parotid Diseases

Introduction

The incidence of congenital facial paralysis has been estimated to be between 0.8 and 1.8 per 1000 births.[1–3] These cases are due to developmental anomalies that occur during embryogenesis. They may present as isolated unilateral or bilateral facial paralysis or as part of a syndrome of other congenital abnormalities, such as congenital unilateral lower lip paralysis or Mobius syndrome.

Case report

A four years and three months old girl was seen initially with a history of suspected hearing loss, recurrent ear infections and hyponasal speech. She had a diagnosis of congenital right-sided facial palsy, which was thought to be slowly improving. There was no history of birth trauma or intra-uterine or peri-natal infection. On examination she had an apparently dense but incomplete right-sided facial palsy. She had weak eye closure but definite facial tone and good movement of the angle of the mouth on smiling. She had symptomatic otitis media with effusion and adenoidal hypertrophy and subsequently underwent adenoidectomy and grommet insertion.

Three and a half years later at a review appointment, she had been unwell and a lump had developed in the right side of her neck. On examination she had a firm mass in the tail of the right parotid. Ultrasound scanning showed a solid mass deep in the parotid with several cervical lymph nodes. Her chest X-ray was normal. She underwent fine needle aspiration of the mass under a general anaesthetic. Cytology suggested origin from lymphoid tissue.

A right parotidectomy was performed via a cervicofacial incision. Exploration revealed a complete absence of the facial nerve; the styloid process and the digastric muscle were identified. A large mass with inflammatory jugulodigastric lymph nodes was excised. The accessory nerve was identified and preserved. A search using a binocular microscope around the periphery of the gland during removal failed to locate any branches of the facial nerve.

Post-operatively facial nerve function was unchanged. Histological examination revealed the mass to be due to non-tuberculous mycobacterial infection. Four months after her surgery the wound was well healed with no evidence of recurrence. Her facial nerve function remains unchanged.

Discussion

Congenital facial paralysis is a rare condition, noticed at birth or soon afterwards. It is important to differentiate congenital facial paralysis from the more common acquired causes of facial palsy such as intra-uterine infection or birth trauma.[4]

Traumatic facial palsy may be associated with difficult and prolonged labour and assisted delivery with forceps. The distinction between congenital facial palsy and acquired facial palsy has important consequences for prognostic, therapeutic and medico-legal reasons. In cases of congenital palsy exploration of the nerve carries no benefit as the nerve is generally represented by a thin fibrous filament or cord peripherally making interposition grafting futile.[5] It appears in this case that the apparent facial movement on the affected side is probably due to neurotization through the orbicularis and the levators.

It is not clear as to whether the main trunk and branches of the facial nerve are completely absent or atrophic and vestigial in cases of congenital facial palsy. It is unlikely that imaging will be of help in differentiating between an absent or atrophic and fibrous facial nerve.

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