The Journal of Laryngology & Otology (2010), ***124***, *1202–1204.*

doi:10.1017/S0022215110001441

Short Communication

Recurrent facial nerve palsy: the role of surgery

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Abstract

Repeated episodes of acute idiopathic facial paralysis present both a diagnostic and a management dilemma. We discuss these issues with reference to four adult cases managed between 1999 and 2008, thus adding to the sparse literature on this subject. For such cases, our unit performs decompression of geniculate, labyrinthine and meatal segments of the facial nerve via a middle fossa approach. A short video, available on *The Journal of Laryngology & Otology* website, demonstrates the key stages of the operation.

We believe there is a role for surgical decompression in cases of recurrent acute facial weakness with three or more documented episodes, especially in the presence of progressive deterioration in function. Decompression of the geniculate, labyrinthine and meatal segments, via a middle fossa approach, is our favoured technique, and has achieved good results.

Key words: Facial Nerve; Decompression, Surgical; Facial Paralysis

Introduction

Recurrent facial nerve palsy represents 4–7 per cent of all cases of acute facial nerve palsy.[1,2] The cause may be idiopathic (i.e. recurrent Bell's palsy) or may be associated with Melkersson–Rosenthal syndrome (a triad of symptoms comprising recurrent facial nerve paralysis, recurrent facial and labial oedema, and fissured tongue).

Recurrent facial nerve palsy may be treated by facial nerve decompression, using a number of techniques.

This article aims (1) to describe a series of four patients who underwent facial nerve decompression via a middle fossa approach for recurrent facial nerve palsy, (2) to present a literature review of the causes of and surgical treatment options for this condition, and (3) to describe our management algorithm for patients with recurrent facial nerve palsy.

A short video also demonstrates the key stages of the operation (Appendix 1).

Case series

Case one

A 27-year-old woman presented with a history of asynchronous, bilateral, recurrent facial nerve palsy. She had suffered a left-sided facial nerve palsy at the age of four years, which had recurred on an annual basis until the age of 14 years. She had also had two episodes of right facial nerve palsy at the ages of nine and 13 years. At the age of 14 years, she had undergone a decompression of the descending segment of the left facial nerve, undertaken by a previous surgeon; pre- and post-operative left facial nerve function had been House–Brackmann grade II. The episodes of left facial nerve palsy had decreased in frequency; at the time of the current presentation, they occurred every three years, with no progressive deterioration in House–Brackmann grade.

At the time of presentation to the senior author RI in 1997, full decompression of the left facial nerve was offered but the patient declined surgery. She continued to have episodes of left facial nerve palsy over the following year; however, these episodes began to be associated with facial swelling. Therefore, a diagnosis of Melkersson–Rosenthal syndrome was made.

At this stage, the patient opted for surgery and underwent total decompression of the left facial nerve via a middle fossa approach.

Within 12 months of the operation, the patient suffered two further episodes of partial left facial nerve palsy, each resolving within 24 hours. Over 10 years' post-operative follow up, she reported no further episodes.

Case two

A 33-year-old woman presented in 2001 with a history of recurrent right facial palsy. The first episode had occurred at the age of 30 years, with complete paralysis lasting two weeks, followed by complete resolution. Two subsequent episodes of partial paresis had occurred, lasting two weeks and two months.

The patient underwent a right middle fossa facial nerve decompression.

Over six years' post-operative follow up, the patient had no further episodes of facial nerve palsy, and remained at House–Brackmann grade I.

Case three

A 42-year-old man was referred in 2002 with a history of 10 episodes of left facial nerve palsy over an eight-year period.

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Presented orally at the XI Facial Nerve Symposium, Rome 25–28 April 2009, Rome, Italy, and at the Politzer Meeting, London 3–5 September 2009, London, UK.
Accepted for publication: 15 February 2010. First published online 17 June 2010.

Each episode had lasted one month, and had involved progressive deterioration in House–Brackmann grade.

The patient underwent a middle fossa left facial nerve decompression. Pre-operative and post-operative function was House–Brackmann grade II.

Post-operative follow up over six years revealed no further episodes of facial nerve palsy.

Case four

A 16-year-old woman was referred in 2008 with a history of complete left facial nerve palsy at the age of 11 years, which lasted one month with complete resolution. Subsequently, she had suffered at least 20 further episodes of left facial nerve palsy, each lasting a week, with progressive deterioration in function to House–Brackmann grade II.

In the pre-operative period, the patient suffered an episode of facial nerve palsy almost every fortnight.

In December 2008, the patient underwent a middle fossa left facial nerve decompression.

Post-operative follow up to date (nine months) revealed no further episodes of facial nerve palsy.

All the patients in our case series underwent pre-operative computed tomography and magnetic resonance image scanning of the brain and temporal bones, assessing the entire course of the facial nerve, to exclude central and peripheral causes of their symptoms. In addition, pre- and post-operative audiometry was also undertaken, and remained unchanged in all patients.

Discussion

Recurrent facial palsy may be idiopathic (i.e. recurrent Bell's palsy) or may be associated with Melkersson–Rosenthal syndrome. Of course, it may also be a presenting feature of underlying pathology, such as a facial nerve tumour or temporal bone dysplasia, which should be excluded by appropriate imaging.[3] Cases have also been described occurring secondary to barotrauma from diving or flying.[4]

Although there is some evidence that Bell's palsy is viral in origin, the aetiology of idiopathic recurrent facial nerve palsy is unclear. Familial cases have been described, the mode of inheritance being autosomal dominant with variable penetrance.[5] An immune-mediated pathogenesis has been suggested by the detection of elevated immunoglobulin levels in patients' blood and/or cerebrospinal fluid, although there is insufficient evidence for this to be conclusive.[6–8]

Melkersson–Rosenthal syndrome is a triad of symptoms comprising recurrent, alternating facial paralysis, recurrent facial and labial oedema, and fissured tongue. It is an autosomal dominant condition with variable expression.[9] The diagnosis is made by detection of the triad of clinical features, although oligo-symptomatic forms of Melkersson–Rosenthal syndrome have been described in which one of the three main symptoms is missing. The diagnosis of Melkersson–Rosenthal syndrome may be delayed if the patient initially has only one of the three symptoms; the other features may become apparent only in subsequent attacks.

In both recurrent Bell's palsy and Melkersson–Rosenthal syndrome, subsequent attacks may be unilateral, contralateral or even bilateral. The literature reports contradictory outcomes following repeated attacks of recurrent Bell's palsy and Melkersson–Rosenthal syndrome. Some reports suggest gradual worsening of facial nerve function, with subsequent residual synkinesis or paresis, especially if the first attack occurs in childhood.[10,11] In contrast, other reports suggest no deterioration in facial nerve function following recurrent episodes of facial nerve palsy.[2]

However, electrophysiological studies have shown loss of action potential amplitude in cases of recurrent facial nerve palsy, compared with cases involving only a single attack, which would theoretically suggest a worse long-term outcome.[12] A proposed aetiology involves neural desynchronisation due to the poor myelination and small diameter fibres of regenerated axons.[13] In addition, the risk of facial nerve palsy recurrence increases with subsequent attacks, ranging from 15 per cent on the second attack to 50 per cent on the fourth recurrence.[2]

These findings imply that surgical intervention may be beneficial, if these factors could be potentially prevented.

The underlying aims of facial nerve decompression in such patients are twofold. The first aim is to relieve the presumed swelling believed to occur during inflammatory episodes, and therefore to prevent nerve function deterioration (i.e. functional intervention). The second aim is to prevent or reduce the number of attacks of facial nerve palsy (i.e. prophylactic intervention).

Some authors have reported partial decompression of the facial nerve via a transmastoid approach, with some success (defined as no further recurrence of facial nerve palsy).[6,14] However, the majority of reports of this surgery describe post-operative recurrence of facial nerve palsy after a short period of remission.[15,16] This is presumably due to oedema in the meatal foramen and labyrinthine segments of the facial nerve.[17]

Decompression of the labyrinthine, geniculate and meatal parts of the facial nerve has also been described (via a middle fossa approach), as has total facial nerve decompression (i.e. also including the horizontal and descending components of the facial nerve) via a combined transmastoid and middle fossa approach. These reports suggest better outcomes for prevention of subsequent facial nerve palsy.[18–20]

Currently, there is no robust evidence to indicate whether surgery reduces the severity and/or frequency of facial nerve paralysis; the literature only gives a number of case series which report their follow-up results. In our unit, we decompress the geniculate, labyrinthine and meatal segments of the facial nerve. Anatomically, this is the narrowest part of the course of the facial nerve, and is presumably where compression occurs when the nerve swells. Our case series results strengthen the evidence that facial nerve decompression, via a middle fossa approach, appears to prevent or reduce the frequency of recurrent facial nerve palsy. We cannot comment on whether this also prevents deterioration in facial nerve function; all our patients had either House–Brackmann grade I or II function pre-operatively, which did not change throughout post-operative follow up.

Our unit offers facial nerve decompression (i.e. meatal, geniculate and labyrinthine segments), via a middle fossa approach, to patients who have had a second recurrence of facial nerve palsy on one or both sides. Authors from our institution have previously described the surgical steps involved.[17]

Recurrent facial nerve palsy is a rare condition; therefore, it is impossible to have a high level of evidence to determine the best management for these patients. The results of our case series add further evidence to the literature. We recommend that patients with a history of recurrent facial nerve palsy should be referred to a skull base centre for consideration of decompression of the facial nerve via a middle fossa approach.

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https://doi.org/10.1017/S0022215110001441 Published online by Cambridge University Press

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Appendix 1. Supplementary video material

A short video, showing the key steps of the operation, is available online at *The Journal of Laryngology & Otology* website, at: http://journals.cambridge.org/sup_S0022215110001441sup001.

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Mr J Doshi takes responsibility for the integrity of the content of the paper.
Competing interests: None declared