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Total decompression of facial nerve for Melkersson-Rosenthal syndrome

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Abstract
Melkersson-Rosenthal syndrome (MRS) is a rare condition characterized by recurrent facial paralysis in addition to various orofacial manifestations. The condition appears to be a granulomatous disorder causing oedema and inflammation of the soft tissues of the face, lips, oral cavity and particularly, the facial nerve. There is general agreement that the symptoms and signs resolve spontaneously, aided perhaps by an empirical course of oral steroids. However, in some patients the condition may be progressive, leading to disfiguring facial synkinesis and increasing residual paralysis. There is anecdotal evidence in the literature to suggest that surgical decompression of the facial nerve in its entire intratemporal course may prevent further attacks of facial paralysis and its sequelae. We present here our experience with surgical decompression of the facial nerve in a 27-year-old woman with MRS who had sufferent recurrent left-sided facial palsy since the age of four. A review of the literature pertaining to facial nerve decompression for Melkersson-Rosenthal syndrome is presented.

Key words: Melkersson-Rosenthal Syndrome; Facial Nerve Surgery

Introduction

Recurrent facial paralysis constitutes less than four per cent of acute idiopathic facial paralyses.[1] Melkersson-Rosenthal Syndrome (MRS) and alternating bilateral palsies are less common causes of recurrent facial paralysis. MRS has been described as a triad of recurrent alternating facial paralysis, recurrent facial and labial oedema (cheilitis granulomatosa) and fissured tongue (lingua plicata).[2]

Facial palsy in MRS occurs in a third of cases and this may be unilateral or bilateral, simultaneous or alternating.[2] The management of facial paralysis in this condition has been a subject of controversy. It is believed that most of the signs and symptoms resolve spontaneously, but, symptoms occur with increasing frequency and persist for longer durations if the condition is untreated. The condition may lead to a disfiguring facial synkinesis and increasing residual paralysis with each episode. The concept of decompression surgery has been particularly appealing to the neurotologist in MRS because the oedematous process evident in the face, lips, cheek and tongue is presumed to be the mechanism underlying the facial palsy. The Michigan group has shown encouraging results with total facial nerve decompression for cases of recurrent facial paralysis including one case of MRS.[3] Nyberg and Fisch[4] have also shown the benefits of middle fossa decompression of the nerve in cases of recurrent facial paralysis.

We present here our experience with surgical decompression of the facial nerve in a 27-year-old woman who had suffered recurrent left-sided facial palsy since the age of four. The disease had been progressive in terms of frequency and duration of acute attacks and the patient also showed signs of synkinesis and residual paresis, that warranted surgical decompression. A review of the literature pertaining to facial nerve decompression for MRS is presented.

Case report

A 27-year-old female presented to the Neurotology Clinic with recurrent facial palsy. The first attack was on the left side at the age of four. She subsequently developed left facial palsies on an annual basis. Each episode involved complete paralysis lasting four to six months, with a near complete to complete recovery. On each occasion she received a course of oral steroids. A tomogram performed at the age of nine showed bilateral dehiscent horizontal parts of the Fallopian canals. By the age of 14, she had undergone all the special investigations to arrive at a diagnosis for her recurrent facial palsies and these were negative. At 14, she underwent decompression of the tympanomastoid segment of the left facial nerve by a transmastoid approach. At surgery, the horizontal segment was noted to be dehiscent being only partially surrounded by cancellous bone. The bone was removed to reveal a large oedematous facial nerve sheath. The nerve was decompressed in both the horizontal and vertical parts with slitting of the nerve sheath. Following this, she had left-sided facial palsies at approximately three-yearly intervals with each episode again lasting for nearly six months with House-Brackmann (H-B) grade 6 palsy.[5] She developed yearly attacks and a residual paresis (H-B 2) over the two years prior to attending the Neurotology Clinic.

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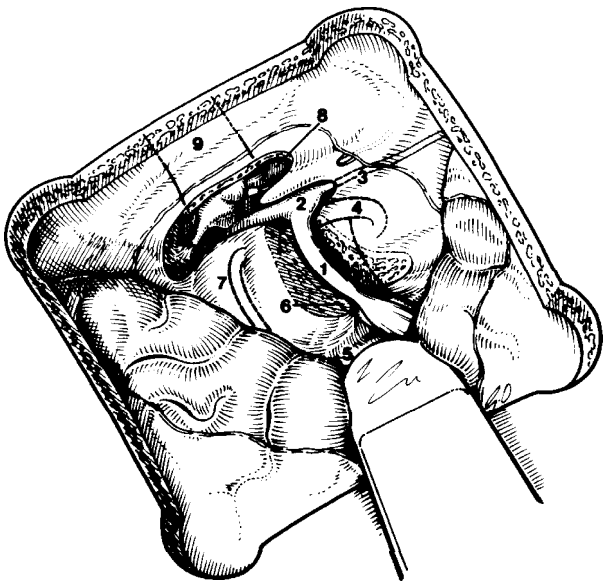

FIG. 1

Schematic diagram illustrating the decompressed labyrinthine segment of the facial nerve in the left middle cranial fossa floor. The important anatomical landmarks in the region are demonstrated: 1 = facial nerve; 2 = geniculate ganglion; 3 = greater superficial petrosal nerve; 4 = cochlea; 5 = superior petrosal sinus; 6 = superior vestibular nerve; 7 = superior semicircular canal; 8 = middle ear; 9 = external auditory canal.

On the right side she had had two episodes of complete facial palsy at the ages of nine and 13, and none since. More lately, she had associated facial swelling and chapping of the lips and tongue, more on the left side than the right. She had complained also of a burning sensation in the lips, tongue and gums for four years prior to presentation. There was no family history of similar complaints. Her hearing was unimpaired and there was no history of vertigo, imbalance, tinnitus or otorrhoea.

On examination, her facial nerve function on the right side was normal and on the left side was a H-B grade 2 with some synkinesis. Computed tomography (CT) and magnetic resonance (MR) images of the temporal bones revealed no abnormality except for the bony dehiscences and previous surgical decompression of the tympanomastoid segment on the left side. A diagnosis of MRS was made and the patient underwent a surgical decompression of the labyrinthine segment of the left facial nerve.

Surgical procedure

A standard temporal 'window' craniotomy was performed, the middle fossa dura elevated and the greater superficial petrosal nerve (GSPN) and the arcuate eminence identified. A small area of tegmen tympani was removed to reveal the tympanic part of the nerve without disturbing the ossicles. The GSPN was followed to the geniculate ganglion and the internal auditory canal (IAC) was identified medially. The labyrinthine segment was then decompressed between the IAC and the tympanic segment as shown in Figure 1. The tegmen defect was covered with bone paté and soft tissue fascia. The temporal plate was replaced and the wound was closed in layers.

Post-operative course

The patient had an uneventful post-operative period and was discharged after five days. In the 12 months since surgery, she has had two episodes of transient facial nerve paresis on the operated side. Each episode lasted less than 24 hours with an associated facial oedema and burning sensation in the oral cavity. Her hearing by audiometry has been normal and there has been no history of vertigo or imbalance. She is delighthed with the results of the surgery and does understand that the operation does not resolve the underlying cause, hence the facial oedema and the non-neurological orofacial manifestations. The patient is on a regular follow-up.

Discussion

Melkersson-Rosenthal syndrome is a rare neuro-mucocutaneous disorder causing localized oedema and inflammation in the face and oral cavity. This syndrome and alternating bilateral palsies are rare causes of recurrent facial paralysis. In MRS, the classic triad of recurrent facial palsy, facio-labial oedema and fissured tongue may not always be seen. Three clinical forms have been described – complete, monosymptomatic and oligosymptomatic. Patients can present with localized cheilitis only and this variant is probably a fairly common cause of constant lip swelling.[6] Ever since Melkersson[7] and Rosenthal[8] first described this condition, nearly 300 cases have been reported in the literature. The prevalence might be higher than the number of published figures, perhaps because the facial nerve is affected in only about 19 to 31 per cent of cases.[2] The disease exhibits a slight female preponderance.[9]

That the condition MRS is an autosomal dominant disorder with variable expression is well established.[10,11] Carr[10] reviewed the familial occurrence of the syndrome and found five families in which two generations were affected and one family with affected individuals in three generations. Male-to-male transmission was observed in one study in a four-generation family.[11] It has been suggested by Smeets *et al.*, that the 'Melkersson-Rosenthal gene' is situated at 9p11 and a case of *de novo* autosomal t(9;21)(p11;p11) translocation has been described.[12]

Granulomatous inflammation is considered a typical histological finding in MRS. Demonstrating a non-caseating epithelioid granuloma by a labial biopsy is mandatory according to some authors,[6] whereas others believe this is not required as MRS is after all, a clinical syndrome.[2] Two distinct types of granulomata have been described by Hornstein.[13] First, tuberculoid-type granulomas appear as tiny, non-caseating epithelioid cell granulomas surrounded by lymphocytes and mononuclear cells with a diffuse oedema of the interstitial connective tissue. Second, lymphonodular-plasmocytic-type granulomas appear as central lymphocytic nodules surrounded by plasma cells and histiocytes in an oedematous connective tissue.

Clinically, MRS presents as a recurrent episodic condition with intermittent periods of remission. The initial orofacial features include – oedema of the face, lips, gingivae, buccal mucosa and tongue; gingival erosions, burning sensation of the tongue and anaesthesia around the mouth and facial palsy. During the course of an attack, which may last from several days to months, the above symptoms and signs may be exaggerated.

The facial nerve is involved in 19 to 31 per cent of the cases.[2] The palsy may initially be a mild paresis but may become complete with progression of the disease. Over several years, the episodes become more frequent and prolonged resulting in distorting sequelae such as facial myokimia and synkinesis. The palsy may also be recurrent simultaneous or alternating, bilaterally.[14] It is not known why the disease affects the facial nerve but spares the remaining cranial nerves. It may be postulated that the inflammation does affect some of the other cranial nerves but subclinically. The facial nerve is the only cranial nerve with a long and tortuous course within a solid bony canal

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TABLE I
CLINICAL FEATURES AND RESULTS OF TOTAL FACIAL NERVE DECOMPRESSION IN MELKERSSON-ROSENTHAL SYNDROME

Authors	Age and sex	Number of palsies	Surgery	Facial nerve function	Follow-up
Graham *et al.*[3]	Not mentioned	8	Total facial nerve decompression	Before surgery – Grade 2 After surgery – Grade 2	No facial palsy in 8 years despite recurring facial oedema
Dutt *et al.* (present study)	27 years (F)	15 (left)	Tympanomastoid decompression (age 14) Middle fossa decompression (age 27)	Before second surgery – Grade 2 After surgery – Grade 2	2 episodes of transient facial paresis (less than 24 hours duration) with facial oedema in 1 year

(Fallopian). This may impede expansion during inflammation to such an extent as to produce a compromise in neuromotor function and hence a facial palsy.

The management of the non-neurological manifestations of the syndrome includes administration of steroids (topical, intralesional or systemic). Other medications that have been found useful are dapsone, clofazimine, sulphasalazine and antihistamines.[2] Limited success has been reported with radiotherapy, sulphazopyridine and reduction cheiloplasty.[15] Systemic and intralesional steroids are perhaps the most effective forms of therapy in controlling progression and reducing some of the orofacial granulomatous oedema.

The surgical management of facial paralysis in this condition is still contentious. Fisch[16,17] has demonstrated encouraging results with surgical decompression of the nerve in its labyrinthine segment in cases of Bell's palsy and herpes zoster oticus. Marsch and Coker[18] noted that anatomical, electrophysiological, radiological, pathological and clinical factors pointed to the meatal segment of the facial nerve as a frequent site of entrapment. They list criteria for considering patients for a prospective surgical trial to assess the role of decompression for idiopathic palsy or herpes zoster oticus which include evoked electromyography (EEMG) studies. However, EEMG does not appear to influence surgical management in cases of recurrent facial paralysis and MRS as the role of decompression is to prevent further attacks and avoid progressive residual paresis.

There have been reports of transmastoid decompression of the nerve with variable results in cases of recurrent facial paralysis and MRS. Canale and Cox[19] performed bilateral transmastoid decompressions of the facial nerve in a case and reported no recurrence of palsy during six years and two-and-half years follow-up. Most other authors performing partial decompression have reported recurrence of paralysis after surgery for MRS and recurrent facial paralysis.[20,21] This was our experience with the present case. Our patient continued to have recurrent palsies after a long period of remission following transmastoid – transmeatal decompression. Presumably, the cause of recurrent palsy in these cases was oedema of the nerve in the meatal foramen and the labyrinthine segment.

Nyberg and Fisch[4] have shown the benefits of middle fossa decompression of the facial nerve in cases of recurrent facial paralysis. A total of 14 patients were operated upon that included 11 patients with acute paralysis in whom electroneuronography (ENoG) demonstrated greater than 90 per cent amplitude reduction within the 10 days following an attack. Three patients underwent middle fossa decompression prophylactically to prevent further recurrences. The Michigan group[3,22] has shown complete resolution with no further episodes in six patients who each underwent a total facial nerve decompression (transmastoid and middle fossa) for recurrent facial paralysis including a case of MRS. Table I shows a detailed account of the characteristics of the two cases of MRS in the literature who have undergone total facial nerve decompression.

The concept of total facial nerve decompression for MRS has evolved logically from the fact that the oedematous process affects the entire nerve within a tight bony canal causing entrapment neuropathy. We recommend a total decompression in cases of MRS where there is a significant increase in frequency, duration and the severity of facial paralysis leading to disabling sequelae such as synkinesis and persistent residual paresis. Total decompression is advised although the labyrinthine and geniculate segments including the meatal foramen appear critical to the success of the surgery. The patients, however, must be made aware that the procedure does not cure the disease process and that the other orofacial manifestations need to be addressed and managed as and when they occur with oral or intralesional steroids.

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