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Idiopathic familial facial nerve paralysis

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Abstract
A 26-year-old man was seen one day after developing a left facial palsy of unknown aetiology. He had previously had a left facial palsy at age 14 and a right facial palsy at 19, both with minimal residual paresis. Both his mother and grandmother have had facial palsies.

The role of hereditary influences in idiopathic facial paralysis, as well as the treatment of this condition, is discussed.

Key words: Facial Paralysis; Family

Introduction

Neumann first described the familial occurrence of facial palsy in 1887.[1] Since then the role of inheritance in the aetiology of idiopathic facial nerve paralysis has been questionned. Familial facial nerve palsy is a rare condition, with a reported incidence between 2.4 and 28.6 per cent of all facial palsies.[1–6]

Recurrence of idiopathic facial nerve paralysis occurs in 2.0–15.0 per cent of the general population,[2,7–9] but in familial facial nerve palsy the recurrence may be as high as 38 per cent, especially if the initial episode occurs during childhood.[1]

We describe here three members of a family with idiopathic facial nerve paralysis, two of whom who have had multiple recurrences.

Case reports

Case 1

A 26-year-old male (Figure 1) presented acutely with a two-day history of left-sided otalgia and facial pain and a one-day history of left-sided facial weakness. His only other complaint was of altered sensation of taste. He admitted to having had two previous episodes of facial weakness at the ages of 14 and 19. His initial palsy had occurred on the left and had resolved spontaneously over two weeks, leaving no residual weakness. His second episode involved the right facial nerve and was treated with oral steroids and physiotherapy. It resolved over one month but left him with slight drooping of the right eyelid and corner of his mouth when he became tired. All three episodes were associated with life stressors such as examinations or work pressure. He had no previous medical history of note and took no medications.

On examination the patient had a grade III House-Brackmann LMN facial palsy on the left and a grade II House-Brackmann palsy on the right. There was altered sensation to light touch in the distribution of the maxillary branch of the trigeminal nerve on the left. No other abnormality was detected in otolaryngological, neurological or other systems. He was normotensive.

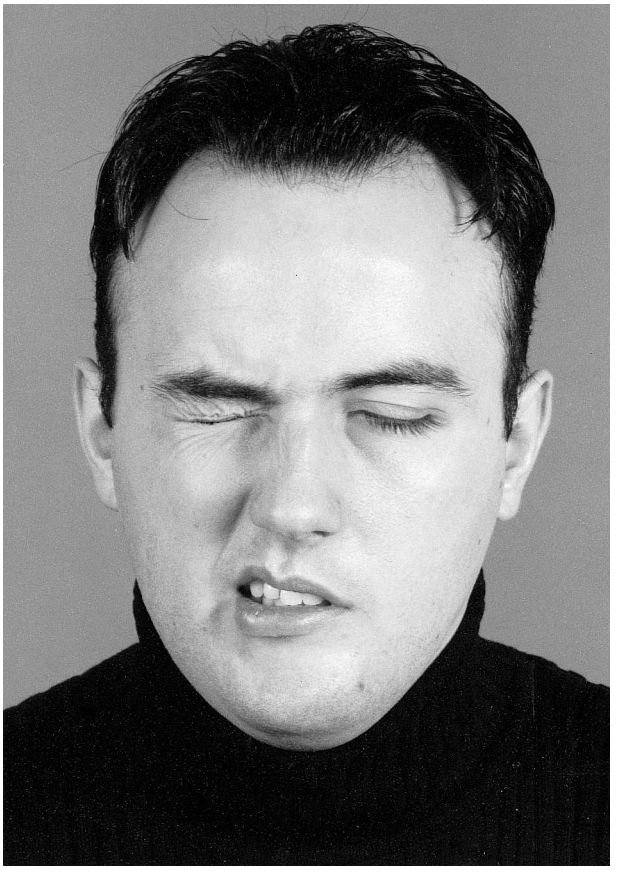

Fig. 1
Left-sided facial paralysis.

Pure-tone audiometry revealed no abnormality, but stapedial reflex testing showed more rapid decay on the left on both ipsilateral and contralateral testing. Urea, electrolytes, liver function tests, thyroid function tests, glucose and calcium were all within the normal range.

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Autoimmune screening, including rheumatoid factor, antinuclear antibodies, smooth muscle antibodies, antireticulin antibodies and mitochondrial antibody screen, were all negative. No rise in titres to either herpes simplex or varicellazoster were detected, nor was IgM detected in response to either of these viruses. However, the herpes simplex titre was consistent with previous exposure to this virus. Further results included an erythrocyte sedimentation rate of 5 mm/hr, a C-reactive protein of less than 10 mg/l, a serum angiotensin-converting enzyme level of 33.0 μ/l and a white cell count of 5.5×10^6/l.

MRI of the brain stem and temporal bones performed 30 days after presentation demonstrated no abnormality. On day two the patient started a 10-day reducing course of prednisolone, diclofenac and cimetidine because of severe left otalgia and neck pain. Physiotherapy was also instituted in the first week. He was reviewed on day 13 and there had been no improvement in his pain. He was initiated on Temazepam, which helped markedly. Sensation over the left maxillary area had returned. On review at day 30, the pain had resolved. Left facial palsy was graded as grade II on the House-Brackmann scale. There was some drooping of the lower eyelid with incomplete closure, but corneal cover was maintained. His sense of taste remains altered.

Questioning revealed a family history as shown in Figure 2.

Case 2

This is the mother of Case 1. She had had asingle episode of right-sided facial weakness at 30 years of age which lasted for three weeks and left no residual weakness. She denied any associated otological problems and felt the weakness had been precipitated by draught. No abnormality was detected on examination.

Case 3

This is the grandmother of Case 1. She has had four episodes of right-sided facial weakness at ages 14, 33, 61 and 70. Each was associated with decreasing facial nerve function. On examination she had a grade III House-Brackmann right facial nerve palsy. No other abnormality was detected on examination.

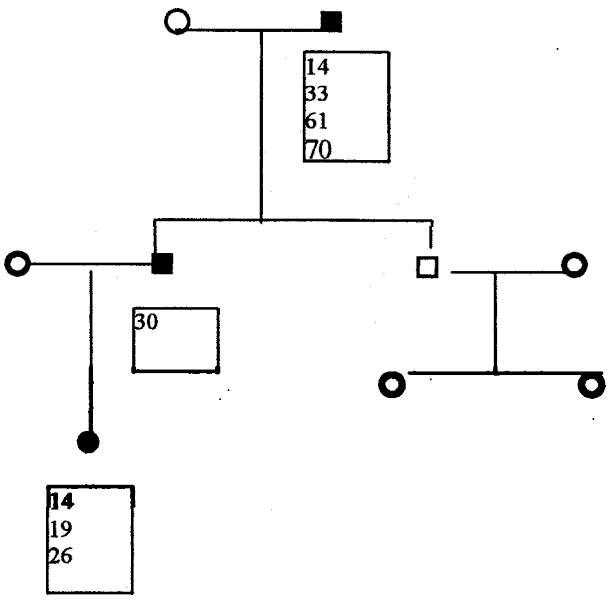

FIG. 2

A family tree detailing the incidence and frequency of facial paralysis. The numbers in the boxes indicate the ages at which the palsies occurred. ○ = unaffected male, ● = affected male, □ = unaffected female, ■ = affected female.

Discussion

Bell's palsy is common, with an incidence of 13–52 cases per 100 000 per annum.[9–11] Idiopathic familial facial paralysis (IFFP), however, is rare. The incidence of patients with a positive family history varies between authors. Familial incidences have been reported at 2.4 per cent,[5] 4.0 per cent,[4] 6.0 per cent,[6] 8.0 per cent,[2] 14 per cent[11] and 28.6 per cent.[3] It has been suggested that in IFFP there is an increased risk of recurrence if the initial episode occurs during childhood. The incidence of recurrence of facial paralysis in the general population is between 2.6 and 15 per cent.[2,4,7–9] Auerbach *et al.*[1] found the recurrence rate to be 38 per cent if the initial episode occurred in the paediatric age group. In their review they identified 10 families with IFFP, 71 individuals were affected, with 13 of these suffering recurrences. This gives an overall recurrence rate of 18 per cent. However, one family accounted for 29 cases, all adults and no member of this family suffered a recurrence. If this family is excluded, then the recurrence rate for all ages is 33 per cent. This leads to the question as to whether or not the risk of recurrence is inherited. Abrol and Maru,[12] in their review of 500 cases of idiopathic facial nerve paralysis, identified 15 cases of recurrence and five cases with a positive family history. In two of these families three family members had each had recurrent palsies, whereas in the remaining three families four members of each had had a single episode of facial palsy. Willbrand *et al.*[6] in their review of 230 patients with idiopathic facial paralysis, found only one case of recurrent paralysis in 14 patients with a positive family history. They did not ask about recurrence in other family members. Takahashi and Fujiwara,[5] in a review of 288 patients, identified seven families with 22 affected members, of whom only four had had recurrences. Of the four, two were from the same family, which had in total five members affected. Overall the evidence is inconclusive, but there do appear to be families in which the risk of recurrence is inherited. The incidence of recurrence appears to be higher if the initial episode occurs in childhood. This may, however, be due to a reporting bias, with multiple recurrences being more memorable to the patient and their relatives, as well as more worrying to parents, thereby increasing their probability of specialist referral, and subsequently publication, especially if the cases are familial.

The mode of inheritance in IFFP is one of autosomal dominance with variable penetrance,[1,3,5,6] as is seen with the family we report. Other forms of facial paralysis that can be inherited in an autosomal dominant fashion, and which must be excluded in the differential diagnosis, include Melkersson-Rosenthal syndrome (MRS), Moebius syndrome, Charcot-Marie-Tooth disease and hereditary neuropathy with liability to pressure palsy (HNPP).

The cause or causes of IFFP remain unknown. In Bell's palsy it has been postulated that it may be caused by a triggering event, such as a non-specific stress-related factor, or a specific infection with an adenovirus or superinfection by a heterotrophic virus which induces the activation of a latent virus present within the geniculate ganglion of the facial nerve.[13–15] There is a growing body of evidence pointing towards this being the herpes simplex virus.[15] Whether or not this plays a role in IFFP is yet to be established. There was no IgM or rise in titres to herpes simplex or varicella zoster detected in this case. However,

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this is not unusual in idiopathic facial palsy and does not exclude a reactivation of latent herpes simplex in the geniculate ganglion.[15] Stress has also been implicated in the aetiology of MRS,[16] and was reported as a recurrent trigger factor by one of our patients. Some authors have suggested that familial facial palsy may be an incomplete expression of MRS because monosymptomatic forms that include only single components of the triad have been described. The other factors that support this are that MRS shows an inheritance pattern, which is autosomal dominant with variable penetrance,[16] and that attacks can have the associated symptoms of headache, trigeminal neuralgiform attacks and a variety of other cranial nerve dysfunctions. The inheritance pattern is consistent with IFFP, as are the polyneuropathy and headache.

Idiopathic facial palsies have been associated with hypertension, diabetes, sarcoidosis and pregnancy. None of our cases was associated with these. Our one case that was examined acutely had initial involvement of the ipsilateral maxillary branch of the trigeminal nerve. Adour *et al.*[2] found that 66 per cent of patients with Bell's palsy had an associated cranial polyneuritis, with the trigeminal and hypoglossal nerves being most commonly involved. They commented that 48 per cent of patients had associated hyperaesthesia in one or more branches of the trigeminal nerve.

Recurrent IFFP remains a particularly difficult entry to treat as the incidence, frequency and severity of the recurrences are highly variable. Treatment therefore remains expectant. Steroids and aciclovir can be used, but there is no evidence to support their use. In this case we used a reducing course of prednisolone because of the severity of pain experienced, in the hope that decreasing any oedema in the facial canal would reduce the pain. It did not appear to have any effect. Surgical decompression has been reported in both recurrent Bell's palsy and MRS, and has been recommended by some authors in all patients with complete facial palsies. Graham and Kemink[17] used a combined transmastoid, middle cranial fossa approach, in two cases of recurrent facial paralysis associated with MRS, with good results. Their reasoning for using the combined approach was that in 94 per cent of Bell's palsies the site of pathologic constriction of the facial nerve is in the intralabyrinthine segment of the nerve. They also commented that other authors had found recurrences of facial palsies after partial decompression. The concluded that caution should be used in recommending surgical decompression until further experience has been gained. With the unpredictability and variability of severity of recurrence in IFFP this option remains experimental.

Conclusion

This case report demonstrates a family with idiopathic facial nerve paralysis in three generations. The pattern of inheritance is consistent with one of autosomal dominance with variable penetrance. The incidence of recurrence in this family is particularly high. The incidence of recurrence appears to be higher in familial facial nerve paralysis if the initial palsy occurs in childhood. There does appear to be a pattern in families with recurrences of idiopathic familial facial paralysis that the increased risk of recurrence is also inherited.

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