

Familial recurrent Bell's palsy

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Abstract

Bell's palsy is a peripheral facial palsy of sudden onset. The etiology of Bell's palsy is unknown and hereditary components may play a role in familial recurrent Bell's palsy. We report three families in which eight patients had a total of 12 episodes of typical Bell's palsy. The pathophysiology of familial recurrent Bell's palsy is discussed.

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Introduction

Bell's palsy, or idiopathic facial paralysis (IFP), is the most common cause of unilateral, lower motor facial palsy. The etiology of IFP remains uncertain. Since the familial occurrence first described in 1887, hereditary factors have been considered to play a role in the etiology of the disease.^[1,2] Here we report three families in whom eight patients have had a total of 12 episodes of Bell's palsy.

Case Report

Family one [Figure 1a]: Four family members had typical Bell's palsy. The proband, a 51-year-old male, presented with an acute right facial muscle weakness and right-sided facial and tongue numbness of one day duration. He had an episode of right facial weakness one year back which resolved after treatment with oral prednisone. On examination he had right lower motor facial weakness with Bell's phenomenon. One of his brothers had two episodes at the age of 32 years and 45 years, his sister had a single episode at the age of 51 years, and his father, who was deceased, had an episode at the age of 40 years. In all of them the facial weakness resolved within a period of one month and the details of the treatment were not known.

Family two [Figure 1b]: Two family members had typical Bell's palsy. The proband, a previously fit 53-year-old male, had two episodes on the left side of the face at ages

36 and 53. Neurological examination revealed left lower motor facial weakness with Bell's phenomenon. He was not given antiviral drugs or steroids and both the episodes resolved completely within a month. His sister had a single episode at the age of 30 years, which spontaneously resolved without any residua.

Family three [Figure 1c]: Two family members had typical Bell's palsy. The proband, a 49-year-old male, presented with acute onset left facial weakness. No history of similar episode in the past. On examination, he had left lower motor facial weakness with Bell's phenomenon. He was treated with artificial tears and methylcobalamin. He refused to take oral prednisone after he was made aware of the possible side-effects.^[3]

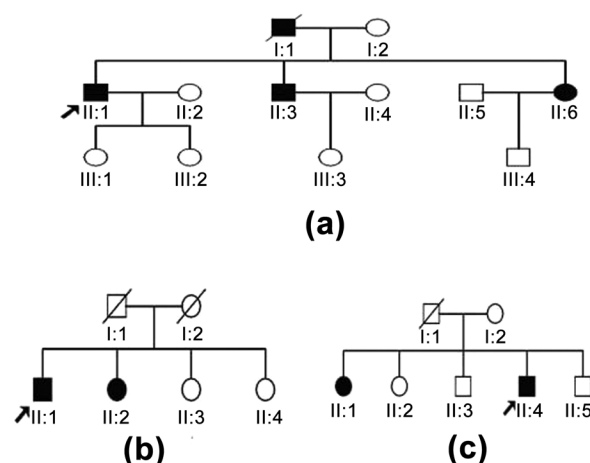


Figure 1a-c: Pedigrees of three families with familial Bell's palsy

His sister had two episodes at the age of 26 years and 40 years. Both episodes resolved completely. However, the details of her treatment were not known.

Discussion

Bell's palsy, which accounts for 60-80% of all cases of peripheral facial palsy, has an incidence of 13-52 cases per 100,000 per year. A familial tendency has been reported in 2.4-28.6% of the cases.^[3] Patients with a positive family history of Bell's palsy were reported to have recurrences more frequently.^[2,4] In this small sample of eight patients with familial Bell's palsy, four patients had recurrences. Recurrent facial palsy can occur in neurological disorders such as Melkersson-Rosenthal syndrome (MRS), Moebius syndrome, Charcot-Marie-Tooth disease and hereditary neuropathy with liability to pressure palsy (HNPP).^[3] These conditions however, have additional features that distinguish them from idiopathic familial Bell's palsy. None of our patients have any symptoms indicative of such diseases.

The exact etiology of Bell's palsy is still uncertain. Since the first report of reactivation of herpes simplex virus (HSV) as the possible cause Bell's palsy in 1972, there has been a growing body of evidence to support this theory.^[5] However, viral etiology is not incriminated in the pathogenesis of familial Bell's palsy. Several possible causes have been proposed for familial Bell's palsy and they include: 1) inherited anatomical abnormality;^[6] 2) vascular risk factors (e.g., diabetes or glucose intolerance and hypertension);^[4,7] and 3) immunogenetic factors.^[8] Of these factors, inherited anatomical abnormality of the facial canal is the most commonly proposed cause for familial Bell's palsy.^[6,9] In patients with no good recovery within three months, further investigations, especially radiological investigations may be warranted to exclude facial canal abnormalities.^[10]

The proposed mode of inheritance in familial Bell's palsy is autosomal dominant inheritance with variable penetrance.^[3] The mode of inheritance in the three families

was suggestive of an autosomal dominant pattern in the family one and an autosomal recessive pattern or autosomal dominant pattern with low penetrance in the families two and three. The association between Human Leukocyte Antigen (HLA) and familial Bell's palsy is still not clear. Some studies showed association between HLA A-31-Cw3-Bw61-DR4 and familial Bell's palsy and some did not.^[1,11] We postulate that familial Bell's palsy is an automimmune disease with genetic predisposition. Clinicians should be aware that facial nerve palsy can be familial, and should enquire for family history especially in patients with recurrent Bell's palsy.

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