

Case Report

Eight episodes of Bell's Palsy in an elderly male: a rare presentation

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ABSTRACT

Bell's palsy is a common, idiopathic, lower motor neuron paralysis of the facial nerve. Multiple recurrent episodes, however, are rare. Most recurrent facial palsies are usually attributable to specific organic, environmental or familial causes. The highest number of idiopathic recurrences reported is eleven, while that in Asia

is five. We present a unique case of eight relapses of Bell's palsy in an elderly Pakistani male. Extensive work-up failed to reveal a cause for the continued recurrence, which was associated with considerable loss of function over time. (Rawal Med J 2013;38:190-192).

Key Words: Bell's palsy, recurrent, unilateral.

INTRODUCTION

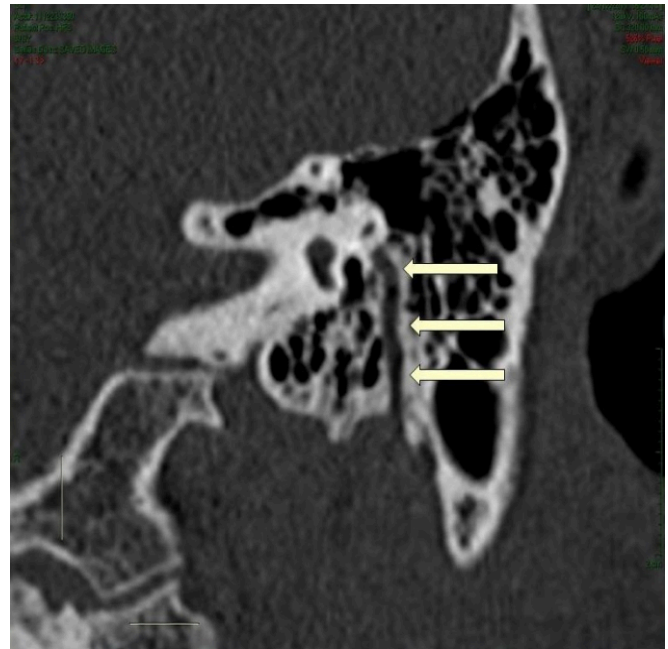
Bell's palsy is an idiopathic lower motor neuron paralysis resulting from dysfunction of the facial nerve. Common symptoms include acute facial palsy, hyperacusis and posterior auricular pain, altered taste sensation over the anterior two-thirds of the tongue and hyperesthesias in the trigeminal nerve distribution. Data shows an incidence of 10 - 30/100,000^{1,2} followed by complete resolution of symptoms within the first two months of onset in 70%80% of cases, leaving 20%30% of patients with varying degrees of residual dysfunction.² While isolated occurrences are quite common, multiple recurrent episodes of unilateral facial paralysis are rare. Most recurrences are usually attributable to specific organic, environmental or familial etiologies leading to the recurring pathology. The highest number of recurrent Bells Palsy reported is eleven,³ while that in Asia is five.^{4,5} In this context, we present a unique case of eight relapses in an elderly Pakistani male.

CASE PRESENTATION

A 71 year-old man presented to our Neurology clinic complaining of the seventh episode of unilateral left-sided facial paralysis in ten years. The first episode occurred at age 60, which was followed by yearly episodes starting at age 65. Being a physician himself, he had self-administered prednisolone tablets during the first five episodes and

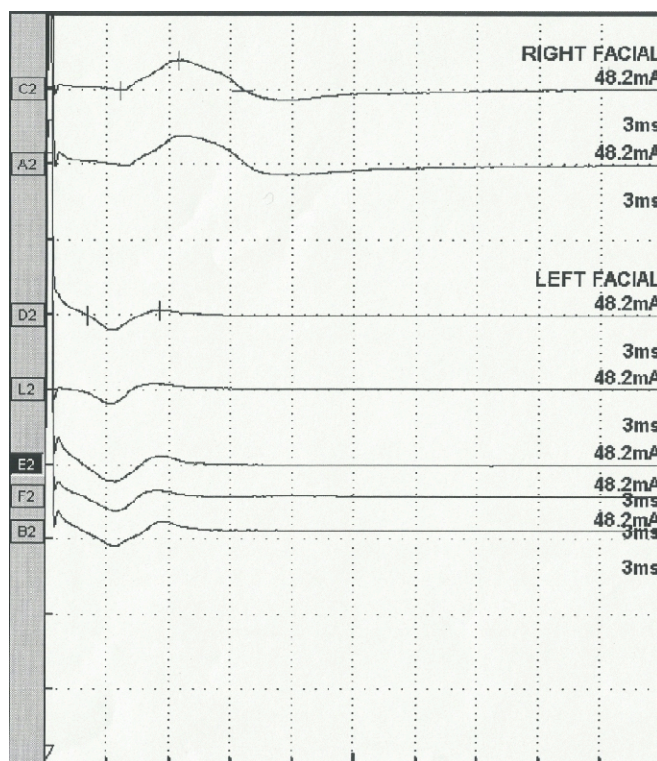
hydrocortisone injections during the last two. Each episode resolved over 2-5 months with subsequent facial weakness of varying degrees in each case. He denied any physical or social limitation due to the episodes; however, the last two were associated with severe pain. He had been diabetic and hypertensive for twenty and ten years, respectively and had previously undergone uvulectomy for recurrent uvulitis.

Fig. 1: High Resolution CT scan section showing normal facial canal (arrows) in the left mastoid.



His presenting episode started ten days prior to consultation. He denied preceding fever or symptoms of an upper respiratory tract infection. He noted that the episodes tended to occur during winter months. Physical examination revealed left-sided lower motor neuron facial nerve palsy corresponding to House-Brackmann Grade IV. No vesicles were noted on the auricle or in the oral cavity. There were no other symptoms to suggest Ramsay-Hunt type II, no family history of recurrent Bells Palsy nor any Melkersson-Rosenthal features such as lingual plicae and labial edema. History was non-suggestive of HIV or Lyme disease. At the time of presentation, he had already electively undergone an MRI scan of the brain, which was unremarkable, ruling out intracranial neoplasm including schwannoma.

Fig. 2: Facial nerve conduction study showing markedly lower CMAP amplitude on the left side.



Rheumatology work up was sought to rule out autoimmune etiologies. Serum ANA, AMA, ASMA and RF were all negative; CRP and ESR were within normal limits. ACE levels were done to exclude

sarcoidosis and was slightly elevated to 57 U/L (normal upper limit=52) on first testing. Chest X-ray was normal. However, given his recent hydrocortisone use at the time, a repeat test was done two months later which was within normal limits. To rule out anatomical causes, a high resolution CT (HRCT) scan of the facial canal was performed and was normal (Fig 1). Herpes IgG was positive.

Four months later, he presented again with the eighth episode, this time accompanied by severe left sided facial weakness. A facial nerve conduction study revealed a 2.07msec onset latency and 0.24mv compound muscle action potential (CMAP) on the left versus 3.66msec and 1.95mv, respectively, on the right (Fig. 2).

DISCUSSION

Approximately 4-7% cases of Bell's palsy experience recurrence.⁶ Most recurrences stop after two episodes and more than four are seldom seen. Yanagihara et al⁷ identified only one patient with more than four recurrences in a group containing 2414 Bells Palsy cases. We identified two cases of five recurrences and one case each of nine and eleven recurrences in the literature. Although evidence of viral and/or environmental triggers was noted in one of these cases, no causal agent could be deciphered in the rest. Associations with pregnancy, malignant hypertension and diabetes have previously been implicated in recurrent Bell's palsy.⁸ However, due to the rarity of >4 episodes, common etiologies such as pregnancy, hypertension and diabetes are unlikely culprits in such patients. If this association was to hold, the incidence of >4 Bell's palsies would have been much higher.

For their patient with eleven episodes (bilateral involvement), Kurca et al postulated an underlying autoimmune cascade, that may be triggered spontaneously or by a multitude of factors including infection by Herpes simplex virus, Epstein-Bar virus or cytomegalovirus, cold weather, insect bite etc.³ In our view, given the very small number of patients who go on to develop multiple, recurrent, idiopathic Bell's Palsy, an autoimmune hypothesis holds promise to explain recurrences versus other

commonly considered etiologies alone (viral, diabetes, hypertension, pregnancy). Select patients may be predisposed to facial nerve inflammation and demyelination secondary to exposure to an inciting antigen leading to cross-reaction with neuronal antigens specific to the intra-canalicular part of the facial nerve. This also explains why Glucocorticoid therapy, combined with antiviral treatment, remains an effective standard of care.⁹ In our case, as well as two others with five episodes each, patients were able to achieve temporary remission on a course of oral prednisolone for one to two weeks. Interestingly, while the literature on Bell's Palsy reports an equal involvement of both sides, we noted a preponderance for left-sided weakness in cases reporting >4 attacks.

While evaluating a patient with recurrent facial palsy, care must be taken to meticulously rule out possible causes before labeling it as an idiopathic paralysis. Multiple recurrences may present as part of a neurological disorder like Melkersson Rosenthal syndrome, Charcot-Marie-Tooth disease or as an autosomal dominant, variable penetrance hereditary Bells Palsy.^{10,11} Recurrences on the same side require evaluation to rule out malignancy particularly schwannoma. Fibrous dysplasia of temporal bone has also been suggested as an etiologic factor for recurrent facial palsy. An abnormally narrow facial canal may be a predisposing genetic factor as well. Finally, despite increasing evidence suggesting autoimmune and viral causes of Bell's palsy, its definitive etiology continues to be elusive for modern medical practice. In the current case, despite a plethora of investigations, we failed to pinpoint a cause for our patient's recurrent Bell's palsy.

Author Contributions:

Conception and design: Arsalan, Khurram
Collection and assembly of data: Arsalan, Khurram, Aasim
Analysis and interpretation of the data: Khurram, Arsalan, Aasim
Drafting of the article: Khurram, Arsalan, Aasim
Critical revision of the article for important intellectual content: Khurram, Arsalan, Aasim
Final approval and guarantor of the article: Arsalan

Conflict of Interest: None declared. This case report was presented as a poster titled "Seven Episodes of Bell's Palsy in an Elderly Male" at the 19th annual conference of Pakistan Society of Neurology. Our patient was yet to have his eighth episode at the time.

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