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Familial Facial Palsy: Another Case Series of Seven Families from the Northern State, Sudan. Is it common?

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Abstract

Idiopathic facial (Bell's) palsy is a common neurologic disease. Familial facial palsy is an uncommon cause of idiopathic facial palsy affecting 4-14% of cases. Most cases follow an autosomal dominant inheritance pattern and many related genes were discovered. One previous study from the Northern State, Sudan reported six families with familial facial palsy. This study aims to describe familial facial palsy in seven families from the Northern State. The total number of cases of facial palsy in this study was 23 with 14 (60.9%) females from seven families compared to 34(38.2% female) cases from six families in the previous study. Two female patients developed recurrent episodes of facial palsy, whereas all other affected patients had a single episode. One of the two females with recurrent facial palsy had 10 episodes of facial palsy. Familial facial palsy may be common in the Northern Sate but further studies are needed to determine the prevalence and incidence in the state and other parts of Sudan. Genetic testing is recommended and family history is important.

Keywords：Bell's palsy, Familial facial palsy, Northern State, Sudan.

Introduction

Sir Charles Bell described the anatomy of the facial nerve and the clinical presentation of facial nerve palsy that bears his name in 1821, although many Greek, Roman and Persian doctors described the facial nerve and facial palsy earlier than Charles Bell [1]. Idiopathic facial palsy or Bell's palsy (BP) is a common neurological disease characterized by sudden onset paresis or paralysis of the facial (seventh cranial) nerve [2]. Bell's palsy accounts for 60-80% of all cases of facial nerve palsy and the annual incidence of BP is about 13–52 cases per 100,000 individuals [3]. No definite underlying cause of facial palsy was identified but many factors may play a role including; viral infections, ischemia, autoimmune disorders, trauma and surgery and the risk is increased in pregnancy, obesity, diabetes mellitus and hypertension [3,4]. Hereditary factors also play a role in the etiology of facial nerve palsy [5].

Familial facial palsy (FFP) is a rare disease characterized by an acute onset of unilateral facial muscle weakness. FFP is not progressive, it resolves spontaneously, and might be recurrent with no obvious precipitating factors [6]. About 4-14% of cases of facial palsy have a positive family history [7]. Most of the reported cases follow an autosomal dominant inheritance pattern but some cases suggest autosomal recessive inheritance and three genes have been implicated [8,9]

Literature review revealed a case series of 34 patients belonging to six different families from the Northern State, Sudan, with single or recurrent episodes of familial facial palsy [10]. We report 7 other families from the Northern State, Sudan, with 23 family members who had single or recurrent episodes of facial palsy.

Patients and methods

This is a descriptive study of the case report type. After obtaining an informed consent, a detailed history was taken from each index case of the seven reported families. The history included personal data, duration, onset and pattern of facial palsy, history of trauma or surgery suggestive of underlying cause, past history , and detailed family history of facial palsy. Thorough clinical examination was performed with special emphasis on the site and type of facial palsy (right/left, upper/lower motor neuron) other neurological deficits and signs suggestive of an underlying cause such as vesicles suggestive of Herpes zoster infection. Family tree was drawn showing the affected family members across different generations.

Case Series

The index case of the first family in this second case series (figure 1) was a 32 years old female from Dongola City, the capital of the Northern State, Sudan. She developed 10 episodes of right side lower motor neuron facial palsy with complete recovery, the first attack was in 2006 and the last attack was on March, 2023, the minimum duration of the episode was two weeks and the maximum duration was 4 months. There was no history of trauma but most of the episodes were preceded by physical or emotional stress such as school or university examinations or loss or illness of her daughter/son or a close relative, one attack occurred during the COVID-19 outbreak. She didn't develop any attack during her three uneventful pregnancies. She is not known to be hypertensive or diabetic. General and neurological examination revealed no abnormality or evidence of an underlying cause of facial palsy. Her mother, who has both hypertension and diabetes mellitus, developed a single episode of right side facial palsy with complete recovery in 2015 but no other family members had facial palsy.

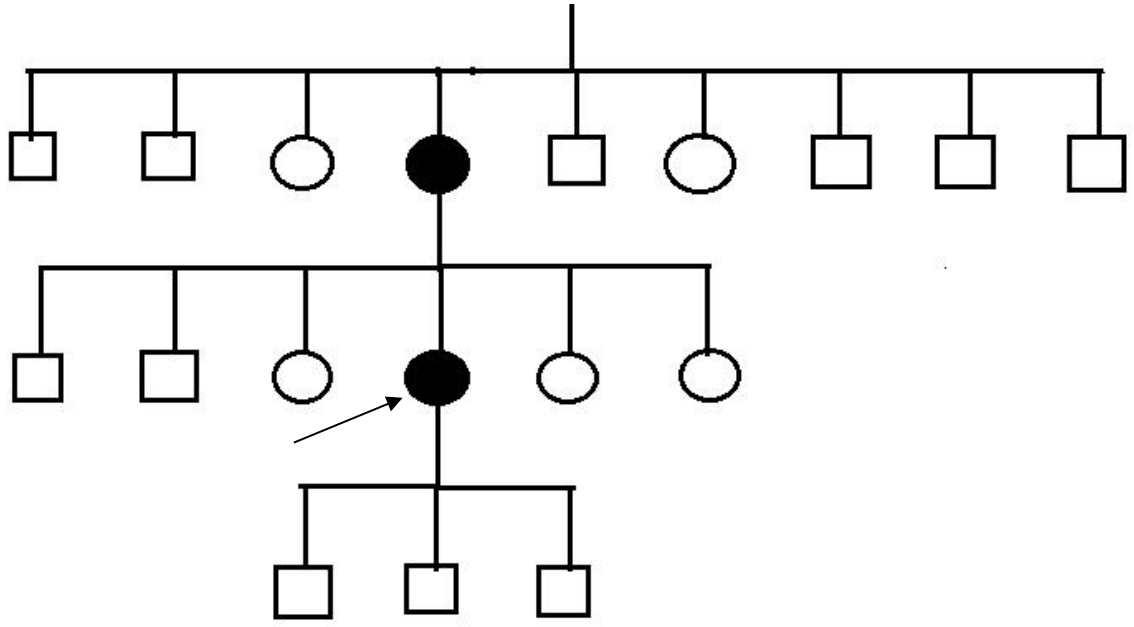

Figure (1): Family pedigree showing two female family members across two generations, with Facial Palsy. The black arrow indicates the index case.

The index case of the second family was a 28 years old female from Al-Selaim, a rural area of Dongola Locality, who presented with acute onset right LMN facial palsy on the 9th of October 2021. She had no past history of similar condition but her family history showed that her 60 years old paternal uncle as well as her 31 years old brother had single episodes of LMN facial palsy as demonstrated in figure (2).

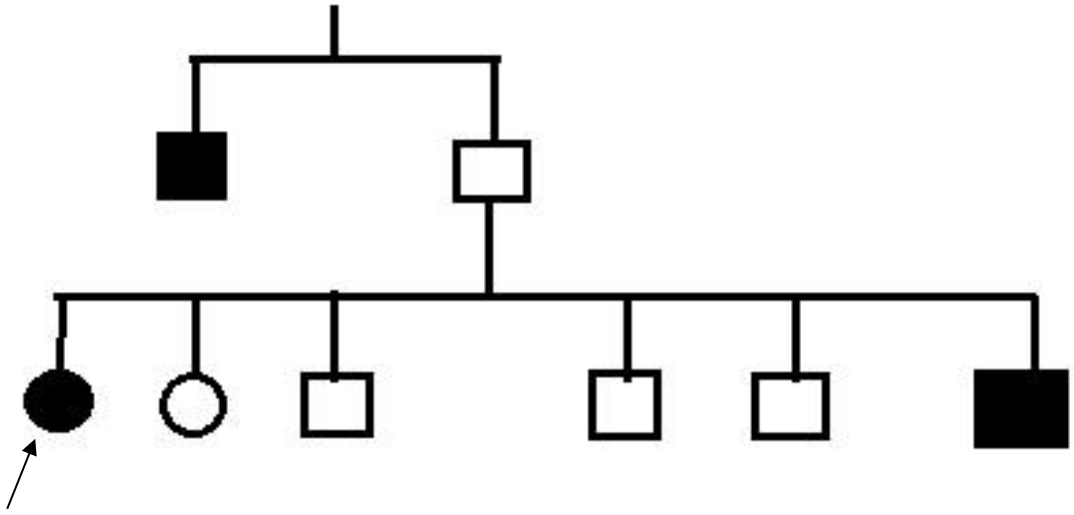

Figure (2): Family pedigree showing 3 family members; two males and a female across two generations, with Facial Palsy. The black arrow indicates the index case.

The index case of the third family was a 55 years old hypertensive female patient from Al-Selaim, Dongola Locality who presented on 20/10/2021 with acute onset LMN facial palsy with no past history of similar condition. Her family history revealed 4 family members, including an eleven years old granddaughter had single episodes of LMN facial palsy as seen on figure (3).

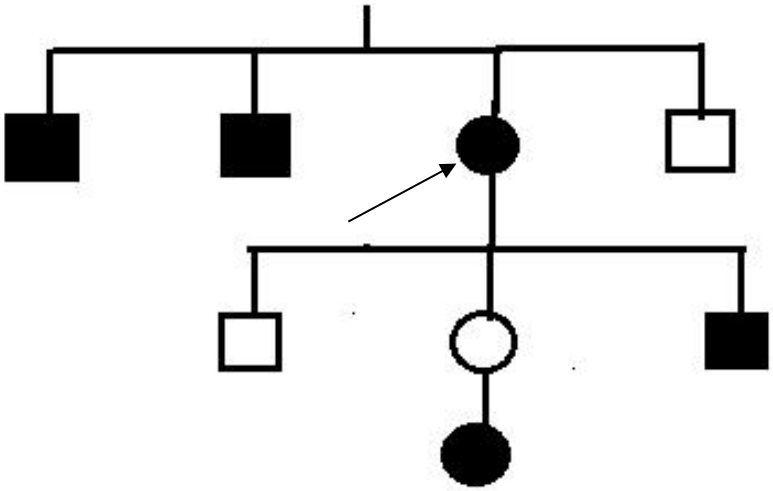

Figure (3): Family pedigree showing 5 family members; three males and two females across three generations, with Facial Palsy. The black arrow indicates the index case.

The 4th family as demonstrated in Figure (4), was from Dalgo Locality and included two patients; the index case was a 29 years old pregnant female with a single episode of acute-

onset right side LMN facial palsy. The patient's paternal uncle had a single episode of LMN facial palsy.

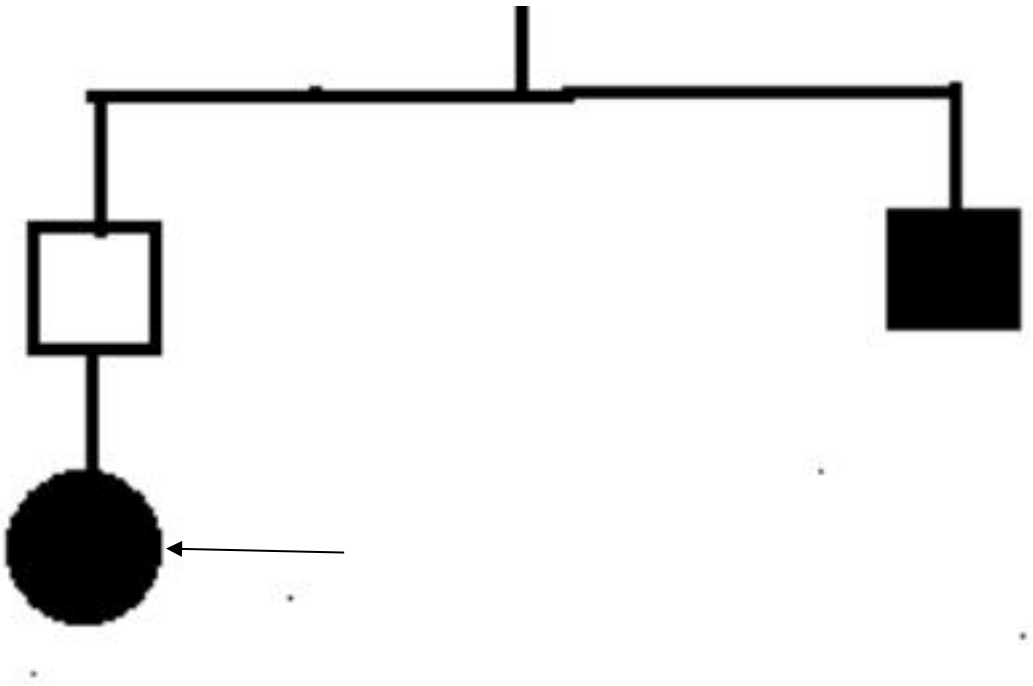

Figure (4): Family pedigree showing two family members; a male and a female across two generations, with Facial Palsy. The black arrow indicates the index case.

The index case of the 5th family was a 65 years old female patient from Akkad, Dongola Locality, known to have both diabetes mellitus and hypertension, who presented with a 5-months' history of right LMN facial palsy with partial recovery. Regarding her family history; two of her sisters, her brother and her first-degree cousin had single episodes of LMN facial palsy as seen in figure (5).

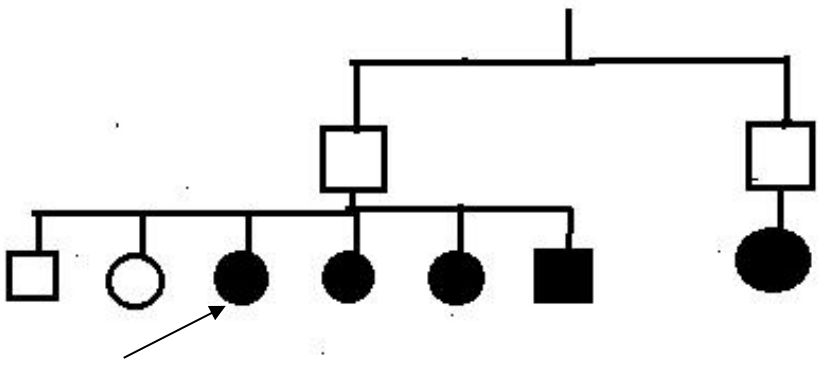

Figure (5): Family pedigree showing 5 family members; 4 females and one male across one generation, with Facial Palsy. The black arrow indicates the index case.

The index case of the 6th family was a 50 years old female with two episodes of right side LMN facial palsy 5 and 3 years prior to presentation with incomplete recovery. Her father had a single episode of LMN facial palsy as shown in figure (6).

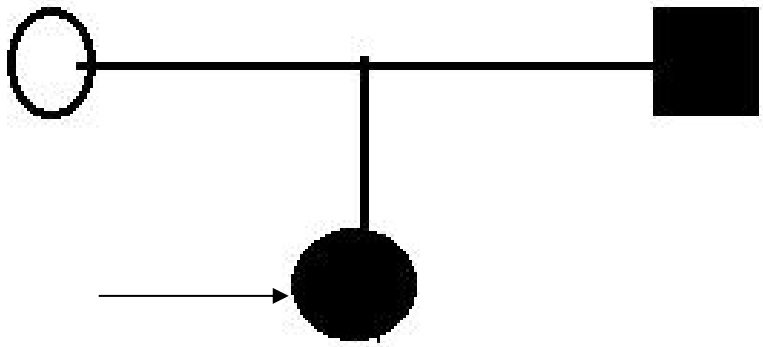

Figure (6): Family pedigree showing two family members; a male and a female across two generations, with Facial Palsy. The black arrow indicates the index case.

The index case of the 7th family was a 40 years old female patient, from Halfa Locality who presented with a two days' history of left LMN facial palsy. Her mother had a single episode at the age of 60 years, her son and daughter had single episodes at the ages of 15 and 20 years respectively as shown in figure (7).

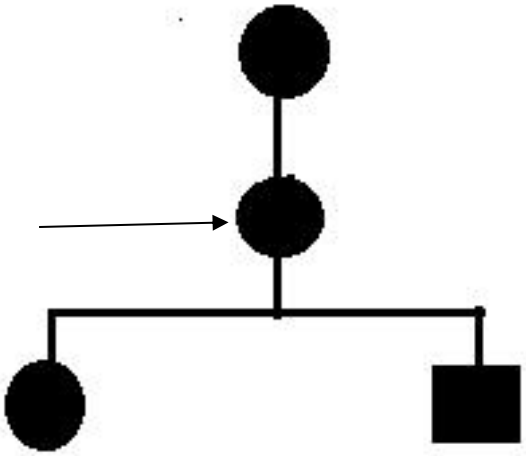

Figure (7): Family pedigree showing 4 family members; three females and one male across three generations, with Facial Palsy. The black arrow indicates the index case.

Discussion

The total number of cases with facial palsy in this second case series was 23 with 14(60.9%) females and a female to male ratio of 1.55:1. The total number of affected cases was less and more females were affected, compared to that reported in the previous study from Northern State which showed that 34 cases from 6 families were affected with 38.2% females [10]. Only two female patients from two different families developed recurrent facial palsy, compared to 7 cases of recurrent facial palsy from 4 families in the previous study[10]. Interestingly, the index case of the first family had 10 episodes of facial palsy whereas that of the 6th family had two attacks, the maximum number of recurrences in the previous study was 3 [10]. There is an 8% increased risk of recurrence after the first episode of facial palsy and the risk increases within familial cases [2]. All the index cases and the other affected family members from the seven reported families did not show any evidence of other underlying causes of facial palsy; such as parotid swelling, trauma, brainstem disease, *Varicella.zoster* infection, or Melkersson-Rosenthal syndrome [11]. The diagnosis was made on clinical background and no neuro-imaging or electrophysiological tests were performed. Genetic testing was not performed due to the high cost and unavailability in the Northern State. All the patients with acute presentation were successfully treated with oral corticosteroids and a few patients needed physiotherapy. The exact cause of facial paralysis in familial cases is not known but many factors might play a role including; narrow facial canal, aplasia or hypoplasia of the facial nerve and immunogenic factors [12,13].

Conclusion and Recommendations

This study reveals that 23 cases of either single or recurrent episodes of facial palsy are reported from seven different families from the Northern State, Sudan, with 14 (60.9%) females and one case with 10 episodes of facial palsy. Considering the number of affected families and the total number of cases in this and the previous study from the same area (57 cases from 13 families); there may be an evidence of increased prevalence of familial facial palsy. Further studies are recommended to determine the prevalence, incidence and different causes of Bell's palsy in the Northern State as well as other parts of Sudan. Genetic testing is also recommended to isolate the implicated genes. We would also like to stress upon the importance of taking detailed family history in all cases of lower motor neuron facial palsy.

Acknowledgement

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Conflict of interest

The author hereby declares that there was no conflict of interest with regards to this study.

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