Sudan Journal of Medical Sciences
Volume 17, Issue no. 1, DOI 10.18502/sjms.v17i1.10690
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Case Report

# Familial Facial Palsy: A Case Series of Six Families from the Northern State, Sudan

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## Abstract

Familial facial palsy is uncommon, accounting only for 4–14% of Bell's palsy cases. We report six families with single or recurrent episodes of familial facial palsy from Northern State, Sudan. The first family had two brothers with single episodes of Bell's palsy. The index case of the second family was a 19-year-old female who and nine other members of her family had a single or recurrent episodes of Bell's palsy. The third, fourth, fifth, and sixth families had eight, five, four, and five members, respectively, who developed either single or recurrent episodes of Bell's palsy. None of the index cases or other members of the six families who were examined showed evidence of facial swelling or fissured tongue suggestive of Melkersson-Rosenthal syndrome. Literature review revealed two studies on Bell's palsy from Sudan but no studies on familial facial palsy. The mode of inheritance was either autosomal dominant with variable penetrance or autosomal recessive. In the second family, there could be a possibility of autosomal recessive inheritance due to increased number of cases after consanguineous marriage. Steroids remain the mainstay of treatment together with protective eye regimens. The role of physiotherapy, although widely used, is controversial. Genetic analysis is recommended and family history should be considered in patients with Bell's palsy.

**Keywords:** Bell's palsy, familial facial palsy, Northern State, Sudan

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Received 04 June 2021
Accepted 05 March 2022
Published 31 March 2022

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Editor-in-Chief:
Prof. Mohammad A. M. Ibnouf

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## 1. Introduction

Idiopathic facial nerve paralysis, also known as Bell's palsy (BP) remains the commonest cause of lower motor neuron (LMN) facial palsy, accounting for about 60–70% of all cases of unilateral facial palsy [1]. Although there are many other causes, familial facial palsy is uncommon and accounts for about 4–14% of cases [2]. Familial facial palsy or hereditary congenital facial palsy (HCFP) presents with different modes of inheritance, with two genes, *HCFP1* and *HCFP2* in cases with autosomal-dominant inheritance and one gene *HCFP3* associated with autosomal recessive inheritance [3]. Melkersson-Rosenthal syndrome (MRS) is a rare disease characterized by the triad of recurrent swelling of various parts of the face such as the upper lip, lower lip, one or both cheeks,

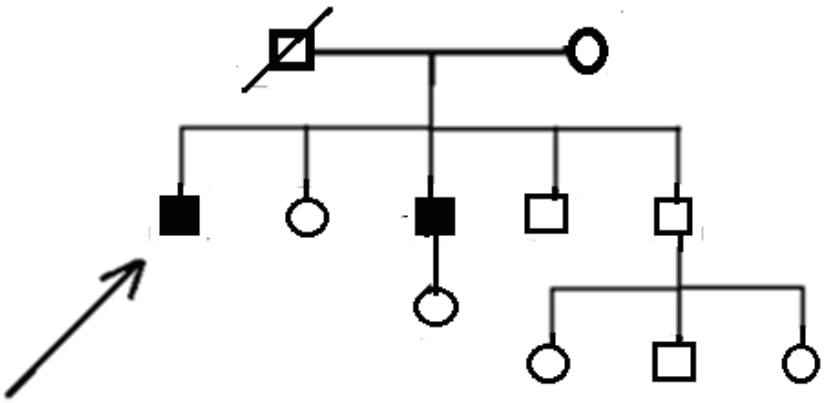

**Figure 1**: Pedigree of the first family from the Northern State, Sudan with familial Bell's palsy affecting two brothers. The arrow points to the index case.

eyelids, or the scalp; recurrent facial palsy; and deep fissures in the tongue. MRS may have a congenital or an acquired etiology [4, 5]. Diagnosis of BP is mainly clinical but investigations, including neurological images and neurophysiological tests, can help identify the underlying cause. Literature review shows two studies on BP in Sudan [6, 7], however, to the best of our knowledge, no study on familial facial palsy has been conducted in Sudan to date. We present six families from Northern State, Sudan with facial palsy and review the family history of similar conditions across different generations of the index cases.

## 2. Case Series

### 2.1. First case

The first family, as seen in Figure 1, included two brothers aged 56 and 48 years from Dongola locality in the Northern State, Sudan who were brought as short cases for the final medical clinical examination at the Faculty of Medicine, University of Dongola in 2016 after receiving informed consent. While the elder brother had right-side LMN facial palsy for about five years before presentation, the younger brother developed left LMN facial palsy for about two years, both had a partial recovery. The two cases showed no evidence of facial swelling or fissured tongues suggestive of MRS, nor did they have herpes zoster or parotid swelling suggestive of an underlying cause of the facial palsy. They had no past history of diabetes, hypertension, or trauma.

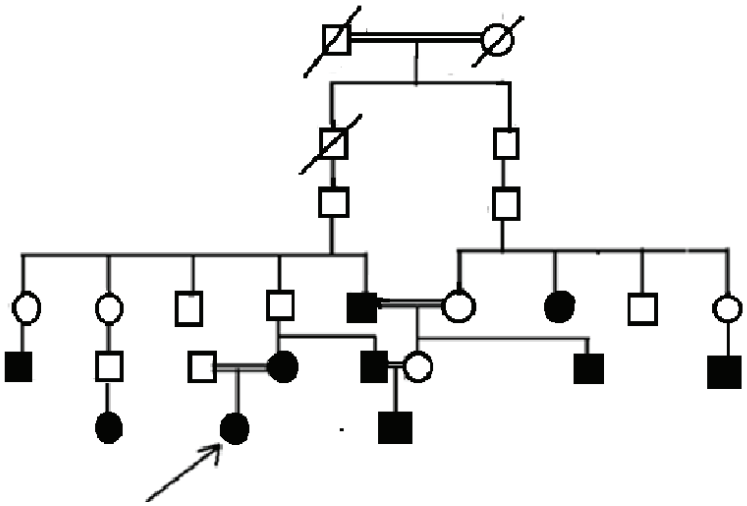

**Figure 2**: Pedigree of the second family from the Northern State, Sudan with familial Bell's palsy with 10 (6 male and 4 female) affected individuals across three generations. The black arrow shows the index case.

## 2.2. Second case

The index case of the second family was a 19-year-old female from a rural area of Dongola locality, who presented to the outpatient clinic of Dongola Specialized Hospital on July 8, 2019 with a single-day history of acute onset deviation of the mouth to the left side associated with drooling of saliva. No headache or loss of consciousness or convulsions, or any evidence of affection of other cranial nerves or limbs weakness or numbness were reported. There was no history of trauma. Clinical examination showed severe LMN right facial palsy with no other neurological deficits, no affection of taste, no hyperacusis, and no blistering eruptions over the external auditory meatus. Examination of the skin and other systems was unremarkable and there was no facial swelling or fissured tongue suggestive of MRS. The patient had a past history of two episodes of left facial palsy with complete recovery, the first at the age of 8 and the second at the age of 10 years. Family history revealed that her mother as well as eight other members of her family had developed either single or multiple episodes of facial palsy as illustrated in the family pedigree in Figure 2. Diagnosis of this patient was done on clinical grounds. The patient was started on oral prednisolone 30 mg daily for a total duration of 10 days with tapering after 5 days, together with lubricant eye drops to avoid exposure keratitis. She was also advised to perform regular facial exercise. Follow-up after one week revealed partial recovery.

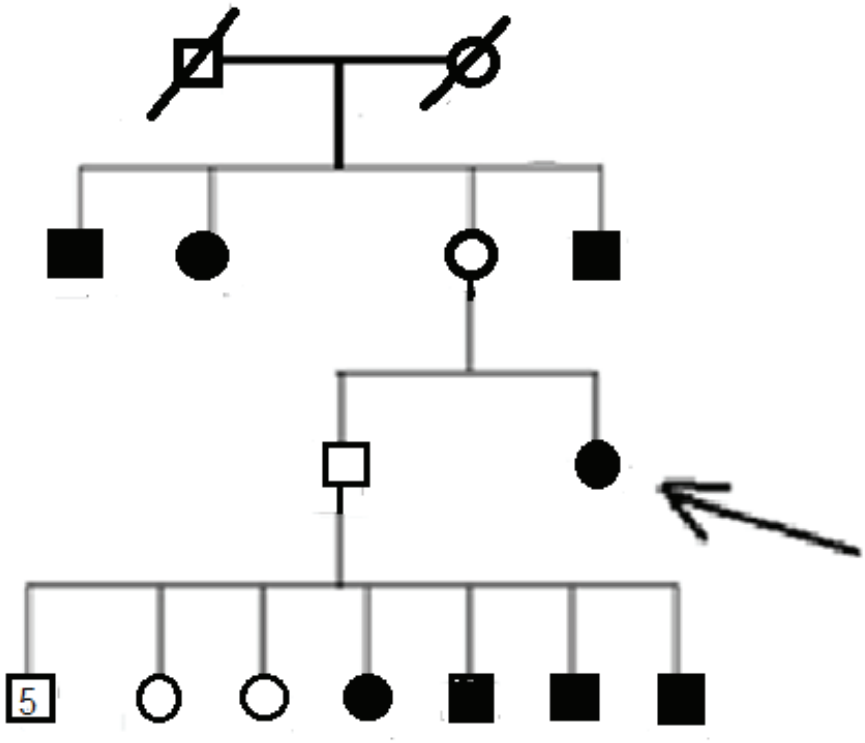


**Figure 3**: Pedigree of the third family from the Northern State, Sudan with familial Bell's palsy with eight (five male and three female) affected individuals across three generations. The black arrow shows the index case. (*Note: The numbers inside the circles and rectangles indicate female and male family members in different generations and their subsequent generations who were unaffected by Bell's palsy.*)

## 2.3. Third case

A 65-year-old female from Wadi Halfa City, Halfa locality, Northern State, who was known to be hypertensive for 10 years but not diabetic, presented on August 20, 2019 with a three-day history of acute onset severe right-side BP with seven other members of her family having single episodes of BP as shown in Figure 3. Examination of the patient revealed no features suggestive of MRS, Varicella-Zoster infection, or any evidence of underlying cause for the facial palsy. She was given prednisolone 40 mg/day as well as lubricant eye drops, follow-up after one week showed partial recovery.

## 2.4. Fourth case

The index case of the fourth family was a 40-year-old male patient from Dalgo locality, who had two episodes of BP; right facial palsy before 17 years of age and left facial at the age of 40. Examination of the patient showed no facial swelling or fissured tongue suggestive of MRS, no blistering skin eruption over the external ear or parotid swelling. There was no past history of trauma. Family history revealed that four other members of

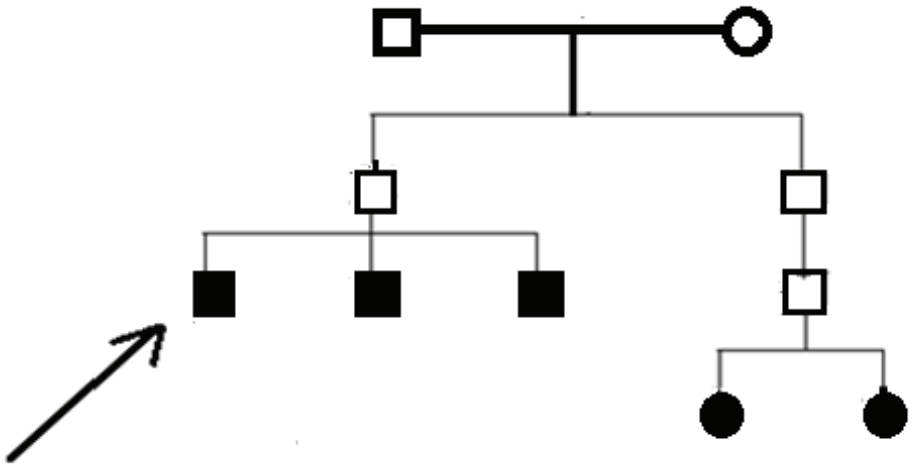

**Figure 4**: Pedigree of the fourth family from the Northern State, Sudan with familial Bell's palsy with five (three male and two female) affected individuals across two generations. The black arrow shows the index case.

his family, including two female children aged three and nine, had Bell's palsy as shown in Figure 4.

## 2.5. Fifth case

The index case of the fifthfamily was a 16-year-old female from Abri City, Halfa locality who presented with right-side LMN facial palsy in 2020 with no features suggestive of MRS or any possible underlying cause. She had a past history of two episodes of similar conditions; the first in 2016 on the left side and the second in 2019 on the right side; both episodes with complete recovery. The family pedigree as seen in Figure 5 shows two of her aunts and one of her uncles with single episodes of LMN facial palsy.

## 2.6. Sixth case

The index case of the sixth family was a 43-year-old male from Dongola locality, who presented with left LMN facial palsy for five days in 2021. There was no swelling of the face or fissured tongue suggestive of MRS. In addition, no blistering eruption suggestive of Varicella-Zoster infection was seen. He had a past history of similar condition on the same side with complete recovery at the age of 13. Family pedigree (Figure 6) revealed that his father, sister, and his 8-year-old son had single episodes of LMN facial palsy while his elder brother had two episodes of Bell's palsy.

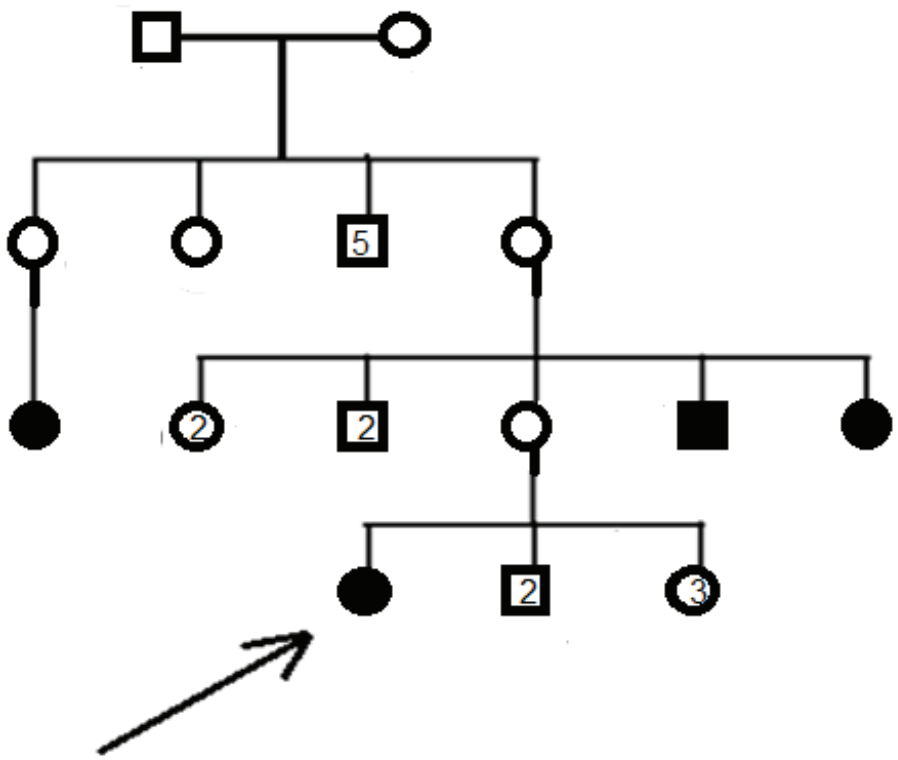


**Figure 5**: Pedigree of the fifth family from the Northern State, Sudan with familial Bell's palsy with four affected individuals (three female and one male) across two generations. The black arrow shows the index case. (*Note: The numbers inside the circles and rectangles indicate female or male family members in different generations and their subsequent generations unaffected by Bell's palsy.*)

## 3. Discussion

Familial facial palsy is uncommon, with few case reports found in the literature [2, 8–10]. Although most case reports suggested an autosomal dominant inheritance with variable penetrance [8–10], the pedigree of the second family may indicate an autosomal recessive inheritance due to the increased number of cases after consanguineous marriage. All affected members of the first and third families had single episodes of BP. The index case and two other members of the second family had recurrent facial palsy whereas the other seven had a single episode of facial palsy. The index case of the fourth family had two episodes of BP whereas the others had single episodes of BP. The index case of the fifthfamily had three episodes whereas the other affected family members had single episodes of Bell's palsy. The index case of the sixthfamily and his elder brother each had two episodes of Bell's palsy. There is no link between the affected families as they come from different parts of the Northern State. Regarding the gender distribution, familial facial palsy affected only males in the first family; males more than females in the second, third, fourth, and sixth families; and females more

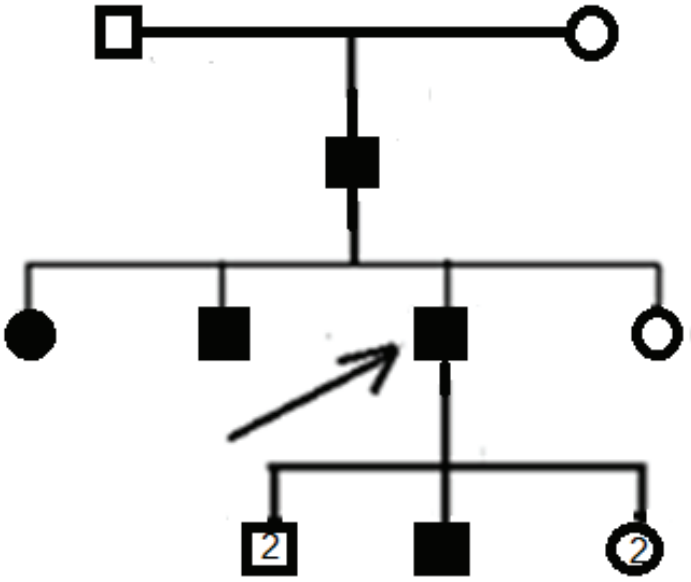


**Figure 6**: Pedigree of the sixth family from the Northern State, Sudan with familial Bell's palsy affecting five individuals (four male and one female) across three generations. The black arrow shows the index case. (*Note: The numbers inside the circles and rectangles indicate female and male family members in different generations and their subsequent generations who were unaffected by Bell's palsy.*)

than males in the fifth family. The index cases of the all six families and other family members with facial palsy who were examined revealed no clinical features suggestive of MRS.

Genetic testing is pertinent for these families to isolate the implicated genes. Our setup and facilities are capable of taking and processing samples of blood or saliva for genetic testing. Eight patients from three of the six families living in or around Dongola city, the capital of the Northern State, with easy access have agreed to perform genetic testing and are ready to give written informed consent for taking their blood or saliva sample for this purpose. Ethical approval to perform genetic testing can be obtained from the Ethics Committee of the State Ministry of Health. Interested individuals or organizations in the field of genetic studies can help us perform genetic testing for these family members.

The underlying cause of familial facial palsy is postulated to be either narrow facial canal, leading to easy compression of the facial nerve, or an autoimmune process involving the nerve [10]. Treatment details of most patients were not known but some of them received oral prednisolone and physiotherapy. Clinical practice guidelines strongly recommend initiation of steroids within 72 hr of the onset of facial palsy as well as

implementing eye care for those with impaired eye closure [11]. The role of physiotherapy is controversial; many methods were adopted including thermotherapy, electrotherapy, massage and facial exercise with variable outcomes and some with complications [12]. Some clinical practice guidelines recommend against physiotherapy [11]. Many grading systems were adopted for evaluation of facial nerve function and assessment of severity and response to treatment of facial paralysis [13].

## 4. Conclusion and Recommendations

In the six families reported here; 2, 10, 8, 5, 4, and 5 family members, respectively, were affected across different generations presenting with either a single episode or recurrent attacks of Bell's palsy. There are a few case reports in the literature with no previous reports from Sudan. Further genetic studies are highly recommended to determine the associated genes. We also stress upon the importance of family history in patients with facial palsy.

## Acknowledgements

The authors would like to thank all the index cases and other members of the six reported families for their consent as well for sharing the detailed history about the disease in their family members.

## Ethical Considerations

Informed consent was obtained from the index cases and the parents of the index case of the fifth family. Any information indicating the identity of the participant such as name, mobile number, or identity documents were excluded for confidentiality purposes.

## Competing Interests

None.

## Availability of Data and Material

All relevant data of this study are available to any interested researchers upon reasonable request to the corresponding author.

## Funding

None.

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