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Case Report

Acute flaccid paralysis in Indonesian adult due to suspected familial hypokalemia paralysis: A rare case

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ABSTRACT

Background: Familial hypokalemic periodic paralysis (FHPP) is rare, so its management is essential to report.
Case presentation: A 25-year-old Indonesian woman complained of feeling weak in both hands and legs, but the heaviest in both legs. The patient has several family members with similar complaints. The patient experienced decreased muscle strength in the upper extremity of 4/4 and the lower extremity of 3/3. Laboratory investigation showed potassium of 2.0 mmol/L, and the patient was given KCL of 50 mEq/24 hours, KSR of 3 × 600 mg/24 hours for 3 days, and a high potassium diet. The next few days, potassium levels increased, and the patient was treated as an outpatient.
Discussion: Early diagnosis and management of acute flaccid paralysis (AFP) due to FHPP are very effective in low-resource settings.
Conclusion: Finding the cause of AFP is essential for better management.

1. Introduction

Hypokalemic periodic paralysis (HPP) is a neuromuscular disease characterized by combined episodes of flaccid paralysis (or muscle weakness) associated with low blood potassium levels [1,2]. It is estimated that as many as 1 : 100,000 people experience HPP, which in women is lower than in men [3,4]. Most cases of HPP are caused by heredity or familiarity, better known as familial hypokalemic periodic paralysis (FHPP). Gene mutations cause FHPP in calcium or sodium ion channels [5]. FHPP is often only identified when a patient's acute flaccid paralysis (AFP) occurs. Based on the description above, we are interested in reporting an Indonesian female with AFP caused by suspected FHPP. We report based on SCARE 2020 guidelines [6].

2. Case presentation

A 25-year-old Indonesian female complained of feeling weak in both hands and legs, but the heaviest in both legs. Daily activities using the extremity feel heavier than the previous days, and no problem related to nutritional intake. As usual, the history of urinating felt complete and not more concentrated. She did not feel any pain or heat. There was no cough, no runny nose, no fever, no nausea, no vomiting, no diarrhoea, and no pounding. The patient had no history of previous illness and no history of using any drug ago. However, the patient's family medical history found similar signs and symptoms (Fig. 1). The patient had never experienced this condition before.

The physical investigation found a muscle strength decrease in the lower and upper extremities [7], muscle strength in the upper extremity at 4/4 and the lower extremity at 3/3. Reflex examination revealed the expected biceps and triceps physiological reflexes, while the patella and Achilles decreased [8]. Laboratory examination showed hypokalemic of 2.0 mmol/L, and others were within normal limits. The electrocardiogram showed what was expected (Fig. 2). The patient was diagnosed with AFP caused by suspected FHPP. The patient received KCL of 50 mEq/24 hours, KSR of 3 × 600 mg/24 hours for 3 days, and a high potassium diet. For 3 days, the patient was monitored for potassium levels. The results were 2.1 mmol/L (the second day) and 3.2 mmol/L (the fourth day). On the fifth day, the patient was treated as an outpatient and given KSR therapy of 3 × 600 mg/24 hours. On the 12th day, the potassium in the patient's blood was 3.8 mmol/L.

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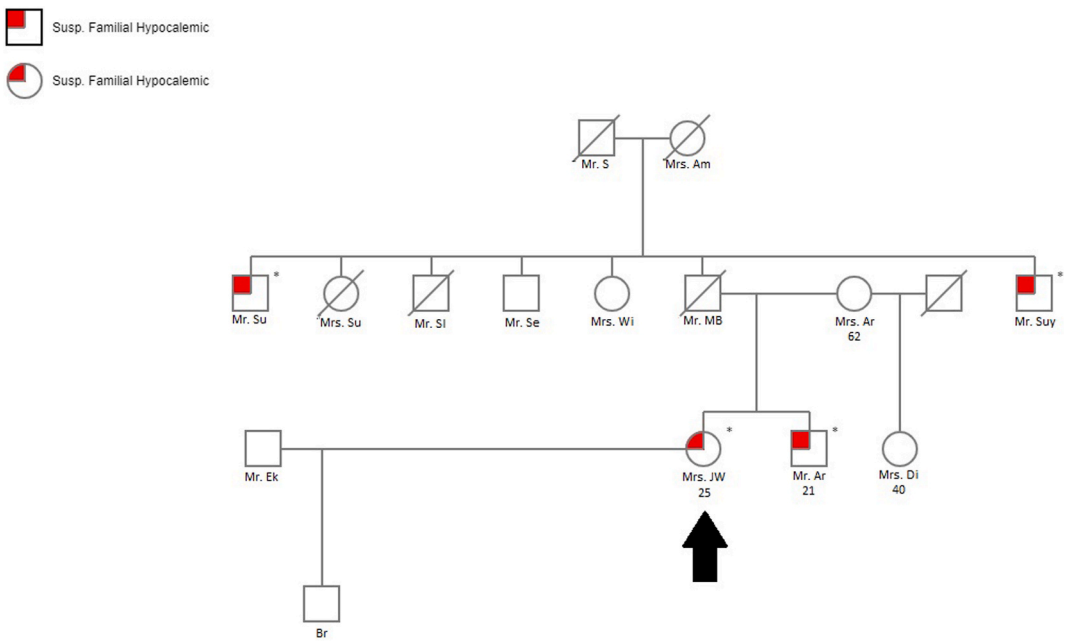

Fig. 1. Medical family genogram.

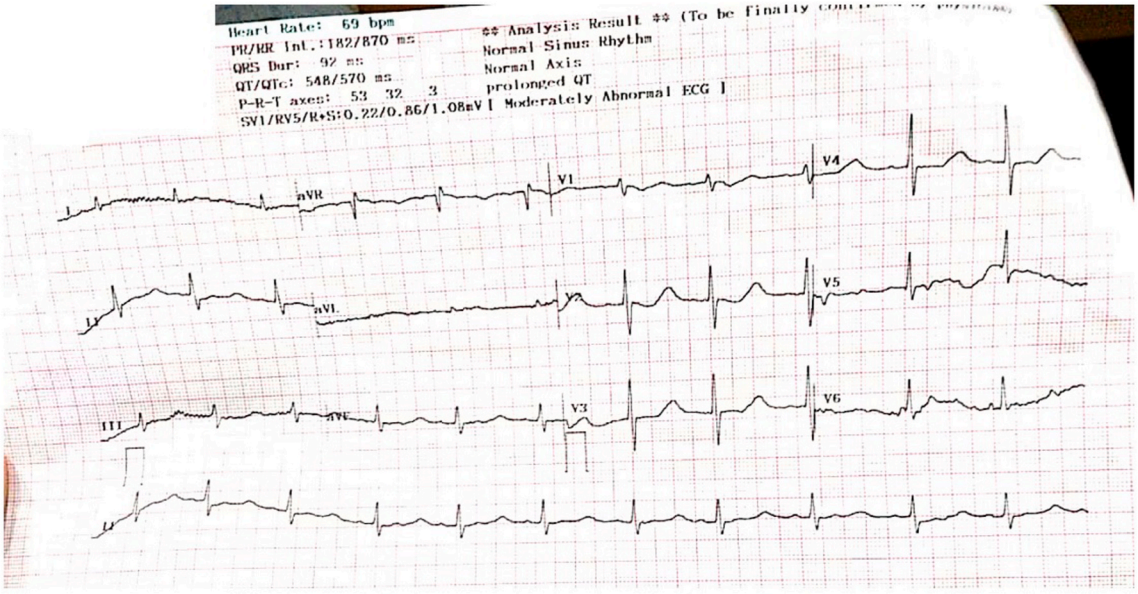

Fig. 2. An electrocardiogram showed a normal sinus rhythm with 92 × /min.

3. Discussion

FHPP is inherited in an autosomal dominant manner with a lower prevalence in women. The pathogenesis of FHPP is associated with mutations in the gene that encodes the ion channel in skeletal muscle, although the exact mechanism is unknown. The most commonly reported gene mutations are those encoding the voltage-dependent L-type calcium channel subunit alpha 1S (CACNA1S), the voltage-gated sodium channel type IV alpha subunit (SCN4A), and the potassium voltage-gated channel subfamily J member 2 (KCNJ2). Several overlapping mutations have also been reported in FHPP, SHPP, and THPP [9].

The basic principles that must be followed when treating patients are controlling plasma potassium, avoiding a lot of glucose and salt load (which results in a transcellular shift), maintaining body temperature, acid-base balance, and careful use of neuromuscular blocking agents. Specific management of hypokalemic FPP is oral potassium supplementation, repeated at 15–30 minutes intervals depending on ECG response, serum potassium level, and muscle strength. Therapy can be administered intravenously if the patient is vomiting or unable to swallow. Prophylaxis against recurrent attacks has been successful with various treatment modalities, including spironolactone and acetazolamide [10].

Potassium administration can be given through oral and parenteral (venous) routes. Oral potassium replacement is safest but is poorly tolerated due to gastric irritation. In mild hypokalemia (potassium 3–3.5 mEq/L), 20 mEq oral KCl can be administered 3–4 times a day, and education of a potassium-rich diet [3,11]. Foods contain sufficient potassium and provide 60 mmol of potassium. Potassium phosphate can be given to patients with combined hypokalemia and hypophosphatemia. In hypokalemia with hypomagnesemia, correction of $Mg2+$ deficiency

should be performed concurrently. The deficit should be corrected gradually over 24–48 hours with regular monitoring of plasma K+ concentrations to avoid transient overrepletion and transient hyperkalemia [3,12]. Effective management of hypokalemic is used to reduce mortality in hypokalemic patients, of which 20% of hypokalemic patients die [13].

The limitation of our study is genetic analysis to confirm the diagnosis of FHPP due to limited resources in developing countries.

4. Conclusion

AFP is a problem that arises due to hypokalemia in the blood. Knowing the etiology of AFP is very important to increase the success of therapy, especially in low-resource settings. AFP due to suspected FHPP is a rare case whose management needs to be reported for better management in the future.

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Ethical approval

This case report does not require any ethical approval.

Consent

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Author contribution

All authors contributed to data analysis, drafting and revising the paper, giving final approval of the version to be published, and agreeing to be accountable for all aspects of the work.

Research registration

Not applicable.

Guarantor

Satriyo Dwi Suryantoro is the person in charge of the publication of our manuscript.

Declaration of competing interest

Devani Bagus Aprinda and Satriyo Dwi Suryantoro declare that they have no conflict of interest.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.104329.

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