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Periodic Paralysis in Children: Case Report and Review of Japanese Literature*)

Yoshikazu NISHI, Kazunari HAYASHI, Ryoji MATSUURA,
Yoshito TANAKA and Tomofusa USUI

Department of Pediatrics, Hiroshima University School of Medicine, Hiroshima 734, Japan

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ABSTRACT

An 8-year-old boy with familial hypokalemic periodic paralysis is described. Twenty eight cases of periodic paralysis under 15 years of age were collected from the Japanese literature and reviewed. Serum potassium concentrations during an attack of paralysis were recorded in 27 cases including our case: 20 cases were hypokalemic, 4 were normokalemic and 3 were hyperkalemic. Hypokalemic periodic paralysis was more frequent in males than in females among adult patients in Europe and the United States, and especially so in Japan and China. Moreover, hyperthyroidism was frequent in Japanese and Chinese adults with hypokalemic periodic paralysis, especially in males, but rare in Europeans and Americans. However, there was no sex difference, and none of these pediatric patients had hyperthyroidism in Japan.

INTRODUCTION

Periodic paralysis usually starts in the second decade of life, and almost all reported patients are adults. To understand the ratio of three types of periodic paralysis (hypokalemic, hyperkalemic and normokalemic) and the complication of thyrotoxicosis in pediatric patients, we present a case of an 8-year-old boy with familial hypokalemic periodic paralysis and briefly review periodic paralysis in children in Japan, where this disease is seen more frequently than in Europe and America.

CASE REPORT

An 8-year-old boy was admitted with the chief complaint of paralysis of the upper and lower extremities. The onset was at midnight, when he awakened with complete flaccidity of all extremities. He was unable to move and had nausea, vomiting, and urinary retention without respiratory distress, irritability, or cloudy sensorium. The attack followed a large high-carbohydrate meal. His 38-year-old father had had hypokalemic periodic paralysis since age 7, usually after a heavy meal. He had been treated with potassium. His thyroid function was normal. None of the other family members were similarly affected and had thyroid disease.

On admission, the patient was motionless, but consciousness was clear. Height was 120.5 cm, and weight 24 kg. The blood pressure was 118/60 mmHg, pulse 60/min. and regular, respiration 32/min. and regular, and temperature 36.5°C. The skin was normal. Movements of the eyes, tongue and facial muscles were intact. The patients was unable to move his neck, shoulders, extremities, fingers or toes. No struma was apparent. The lungs were clear, and the heart sounds were normal. The muscles were extremely hypotonic and weak. No deep tendon reflexes could be elicited.

Routine complete blood count and urinalysis were normal. Liver and renal function tests were normal. Fasting blood sugar was 110 mg/100ml. Thyroid function tests were normal (T_4 7.2 μg/100 ml and TSH 7.1 μU/ml). Serum sodium was 144 mEq/l, chloride 108 mEq/l, potassium 1.7 mEq/l, calcium 9.8 mg/100 ml, and inorganic phosphorus 3.9 mg/100 ml. Plasma zinc concentration was low 65 μg/100

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Table Summary of periodic

Case No.	1	2	3	4	5	6	7	8	9	10	11	12	13	14
Sex	F	F	F	M	M	F	M	M	F	F	F	M	M	M
Age at diagnosis	4y8m	4y3m	2y3m	9y10m	6y	12y	6y2m	15y	1y	11y	10y	15y	14y	6y4m
Age at oneset		1y2m	1y2m	8y		5y	1y6m	12y	10m			12y	3y	8m
Inheritance	+	+	+	−	+	−	−	+	−		−	−		−
Predisposing factor														
heavy meal	+	+	+		+		+	+					+	
vigorous exercise														
Thyroid function					N	N				N			N	N
Complication														
Serum K (mEq/l)		2.7	2.7	↑	4.4	3.3	1.5	↓	3.7	↓	3.5	3.5	↓	6.0

ml (normal: 97.2±21.2 μg/100 ml) and erythrocyte zinc concentration was normal 36.7 μg/gHb (normal: 37.9±4.0 μg/gHb).

Intravenous therapy with 35 mEq KCl in 1,000 ml of normal saline was administered in the next 16 hours (60 ml per hour). Three hours after therapy was begun, the patient could move his lower extremities slightly and was more alert. Thirteen hours later he could move all extremities without difficulty, his hand grasp was strong, and deep tendon reflexes were also active. He could eat and drink well. He was up and moving about normally. No muscular weakness could be detected. Serum potassium at this time was 3.8 mEq/l and intravenous therapy was discontinued. Plasma zinc concentration became normal 85 μg/100 ml and erythrocyte zinc concentration was normal 40.4 μg/gHb.

REVIEW OF THE LITERATURE

A total of 28 patients with periodic paralysis under 15 years of age was collected from the Japanese literature based in a survey of Japan Centra Revuo Medicina extending from 1951 to 1980.

The table lists the pertinent clinical and laboratory for 29 patients, including our case. There is no sex difference (14 males; 15 females). The age at onset was recorded in 22 case; 12 patients had the first attack under 5 years of age. In 12 cases there was family history of muscle weakness in episodic attacks. The ingestion of high carbohydrate meal was considered to be a predisposing factor in 14 patients. Thyroid function was tested in 15 patients and was normal in all 15.

Other disorders were present in 3 patients: potassium wasting nephropathy in case 15, renal disease in case 18, and ectopic pinealoma, panhypopituitarism and diabetes insipidus in case 23. Serum potassium concentrations during the attack of paralysis were recorded in 27 cases. Hypokalemia was noted in 20 cases, normokalemia in 4 and hyperkalemia in 3.

DISCUSSION

Periodic paralysis is a familial disease characterized by recurrent attacks of weakness or paralysis of the somatic musculature, accompanied by loss of deep reflexes and failure of the muscles to respond to electrical stimulation.

Alterations in serum potassium during attacks have led to a differentiation of three types of periodic paralysis: (1) hypokalemic, the most common of the three types; (2) hyperkalemic, also called adynamia episodica hereditaria; and (3) normokalemic, the least common of the three forms. In all types there is a strong familial tendency, and inheritance is usually autosomal dominant although sporadic cases have been reported.

The most consistent factors predisposing to an attack are a heavy carbohydrate meal, prolonged rest after vigorous exercise and exposure to cold[1]. In our study the attacks of weakness occurred after the ingestion of a high carbohydrate meal in 14 cases and after vigorous exercise in 2 cases.

In Europe and the United States, hypokalemic

paralysis under 15 years of age in Japan

15	16	17	18	19	20	21	22	23	24	25	26	27	28	present case
F	F	M	F	M	M	M	M	M	F	F	F	F	F	M
14y	10y	14y	15y	15y	13y	14y	15y	15y	13y	15y	11y	7y	12y	8y
13y	3y			11y	11y	12y	10y		3y	5y	5y	3y	8y	8y
−	−			−	+	+	+		+	−	+	+		+
					+	+	+		+		+	+		+
									+		+			
N		N		N	N	N	N		N	N	N			N
potassium wasting nephropathy			renal disease					ectopic pinealoma panhypopituitarism diabetes insipidus						
2.1	2.5	2.1	↓	5.3	1.6	2.0	2.3	↓	1.8	2.7	4.6		N	1.7

periodic paralysis occurs two or three times more frequent in males than in females[3]. In Japan, almost all patients with this disorder have been males[6]. However, in Japanese pediatric cases no sex diffecence was observed in our study.

Periodic paralysis appears to be a rare complication of hyperthyroidism in Europeans and Americans. However, this disorder is a common complication of thyrotoxicosis in Japanese and Chinese, usually among patients without a positive family history[2,4]. In Japan, among 1,214 male patients with hyperthyroidism, periodic paralysis was observed in 99 (8.2%); and among 4,847 female patients with hyperthyroidism it was observed in 20 (0.4%)[4]. Moreover, approximately half of the patients with hypokalemic periodic paralysis had hyperthyroidism in Japan[5]. Of 1,366 Chinese patients with hyperthyroidism (1,188 females and 178 males), 23 (13%) males and 2 (0.17%) females gave a history of one or more attacks of paralysis[2]. However, of the children with periodic paralysis in our study none had hyperthyroidism.

It is far from fully explained why periodic paralysis is a common complication of thyrotoxicosis in Japanese and Chinese adult but not in pediatric patients and not in Europeans and Americans, and why male adult patients are more numerous than females although there is no sex difference in pediatric patients in Japan.

Within our knowledge, there has been no report on zinc metabolism in periodic paralysis. Plasma and erythrocyte zinc concentrations increased after KCl therapy in our patient. The role of zinc in periodic paralysis is not known, although zinc is a key nutrient which modulates cellular proliferation, division and metabolism such as Na^+-K^+ ATPase[7]. Further studies are needed to clarify these problems.

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