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PROLONGED PARALYSIS AS AN UNUSUAL PRESENTATION OF RENAL TUBULAR ACIDOSIS

A. DERAKHSHAN AND G.H. HASHEMI

From the Division of Pediatric Nephrology, Department of Pediatrics,
Shiraz University of Medical Sciences,
Shiraz, I.R. Iran.

ABSTRACT

A case of renal tubular acidosis (RTA) who had gradual onset of paralysis of lower extremities and persisted for a long time is presented. His primary workups were mistaken for muscular dystrophy. Eventually, an abdominal ultrasonography revealed small stones in both of the kidneys and his workup for the stones were in favour of distal renal tubular acidosis and advanced rickets with hypokalemia. He had a dramatic response to alkaline therapy. There are many reports of periodic paralysis in RTA but no report of prolonged paralysis is found in the literature.

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INTRODUCTION

Renal tubular acidosis was first described by Lightwood[1] et al. and Butler[2] et al. in children and by Baines[3] in adults. On clinical and pathophysiological grounds, RTA is classified into several types.[4] Rickets is not a usual presentation in isolated proximal RTA but accompanied with Fanconi's syndrome and associated phosphaturia, osteomalacia and rickets are usual findings.[5] Although delayed diagnosis of distal RTA in adults and older children causes demineralization of bone, advanced rickets has not been reported, except for vitamin D deficient individuals.[5,6] There are many reports of hypokalemic periodic paralysis, almost all of them in adults, in association with other conditions, such as Wilson's disease,[7]
vanadium poisoning[11] and sporadic form of distal RTA.[12,13]

A case of classic (distal RTA) with advanced rickets, paralysis of lower extremities and dramatic response to treatment in a 10-year-old boy is presented and discussed here.

CASE REPORT

A 10-year-old boy presented with an abnormal abdominal sonography (small stones in both kidneys), which had been done due to vague abdominal pain. He had sufferred from pain and progressive weakness of lower extremities since 18 months before, so that he had not been able to walk for about 15 months. His previous workups were as follows: EMG and NCV once reported as spinal muscular dystrophy and in another occasion, as myopathic process, serum Ca= 9.9 mg/dL, P=2.9 mg/dL, alkaline phosphatase (ALP)= 1707 IU, CPK and LDH were normal.

In another occasion, BUN= 15 mg/dL, P= 2.7 mg/dL, Ca= 9.9 mg/dL, ALP= 1650 IU. Muscle biopsy reported primary muscular dystrophy.

Physical examination: undernourished, well developed child, brought on wheelchair, weight= 24.8 Kg, height= 129 cm, BP= 90/60 mmHg.

Deep tendon reflexes were barely detectable in upper extremities, but absent in lower extremities. Investigations into his renal stone began. Arterial blood gas (ABG): pH= 7.26, HCO_3= 9.4 mEq/lit, PCO_2= 21.7 mmHg, BUN= 10 mg/dL, Cr= 0.6 mg/dL uric acid 2.8 mg/dL, Na= 139 meq/lit, Cl = 120 mEq/lit, K=3 mEq/lit, P= 2.7 mg/dL, Ca= 9.3/dL mg, ALP= 1494 IU, urine Ca/Cr= 0.9, 24 hr urine Ca= 224 mg, urine pH= 7.2, urine Na= 93 mEq/lit, urine K= 26 mEq/lit, urine Cl= 86 mEq/lit, serum anion gap= 9.6, urine anion gap=33 and finally wrist X-ray revealed generalized osteopenia and advanced rickets.

Another kidney sonography and KUB revealed nephrocalcinosis rather than stone. Under the diagnosis of distal RTA, treatment started with polycitra solution and also

Table I. Clinical and paraclinical findings at presentation and 8 years follow up.

Interval (year) / variable	0 *	0.5	1	2	3	4	5	6	7	8
Age (year)	10	10.5	11	12	13	14	15	16	17	18
Weight (kg)	24.9	31	35.9	39.9	51.5	54	58	61.5	65	70
Height (cm)	129	136	142	147	154	160	166	172	174	174
pH	7.26	7.36○	7.38○	7.37*	7.4*	7.34*	7.35*	7.36*	7.4*	7.4*
HCO_3 (mEq/L)	9.4	18.5	21.2	20	20	18.5	20	22	24	24
PCO_2 (mmHg)	21.7	34	36.5	35	33	35	34	37	35	35
Ca (mg/dL)	9.3	8.5	9.5	9.5	0	0	0	9.5	0	9.5
P (mg/dL)	2.7	5.3	5.5	5.1	0	0	0	5.5	0	5.3
ALP (IU)	1494	879	620	0	0	0	0	570	0	380
Na (mEq/L)	139	145	0	0	145	0	0	0	0	140
K (mEq/L)	3	4.2	0	0	4.5	0	0	0	0	4.5
UCa/Cr**	0.9	0.16	0	0.12	0.11	0	0.15	0.05	0.11	0
Polycitra (mEq/kg)	6	6	5	5	4	4	2.5	2.5	2.5	2.5

*at presentation

●venous blood gas

○not done

** Urine calcium / Urine creatinine

600,000 unit vitamin D3 was orally given. He had a dramatic improvement after the initiation of treatment. One week following the treatment, he could walk with help and one month later he could walk normally.

Six months after the beginning of treatment he had an almost normal physical examination. His sequential clinical and paraclinical findings during 8 years follow up are summarized in Table I. Now he is doing well in 2nd year of engineering school with normal kidney function tests on 2.5 meq/kg polycitra solution.

DISCUSSION

There is no doubt about the diagnosis of dRTA in this case with normal anion gap, hyperchloremic metabolic acidosis, alkaline urine pH, positive urine anion gap, hypercalciuria and nephrocalcinosis.[4-6] There are many reports of periodic hypokalemic paralysis in the literature in patients with RTA, singly or in association with other conditions.[7-13]

The problem that caused misdiagnosis in our case was his prolonged paralysis and also the results of muscle biopsy and EMG which were in favor of a myopathic process. A guiding point in his previous lab data not considered was very high alkaline phosphatase and low phosphate level. Paralysis of our patient was most probably due to hypokalemia and hypophosphatemia and associated rickets.[5,6,14] Although acute hypokalemic paralysis is a rare cause of acute weakness, its detection and intensive evaluation of various etiologies require special considerations.[15]

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