Exploratory laparotomy revealed the fistula opening internally at the isthmus of the uterus. At the isthmus the uterus was perforated and fetal remnants in the form of a few long bones were lying outside. A loop of the small bowel was densely adherent at this site which perforated during separation. There was no evidence of regional ileitis or endometriosis. Abdominal hysterectomy and resection and anastomosis of the small bowel was carried out. (The couple had requested sterilization.) The fistulous tract was excised. Histopathology revealed no evidence of endometriosis, Crohn's disease or tuberculosis. Postoperative recovery was uneventful and the external opening healed completely.

Discussion

A fistula is an abnormal communication between two epithelial surfaces. Fistulae are usually lined by

granulation tissue but they can become epithelialized. Most fistulae originate from trauma or from some type of infectious process that disrupts the continuity of the tissues involved. Once a fistula is diagnosed the basic principle in the treatment is obliteration of the primary opening of fistulous tract. There is no non-surgical treatment for fistula.⁴

In this case the patient possibly developed uterine perforation leading to pelvic abscess extending to perinephric and right flank region due to septic abortion which were drained subsequently. Conceivably the uterine perforation with stenosis of the cervix due to interference led to communication with the right flank and drainage through this area gave rise to the uterocutaneous fistula. Injection of contrast through the fistulous opening permitted an accurate diagnosis and extirpation of the uterus produced a cure.

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Type IV renal tubular acidosis associated with Alport's syndrome

Ružena Tkáčová, R. Roland, A. Böör¹, Anna Kováčová², Ivica Lazúrová³, I. Tkáč⁴, T. Hildebrand and P. Šefara

*Department of Internal Medicine I, and Departments of*¹*Pathological Anatomy,*²*Biochemistry,*³*Internal Medicine II, and*⁴*Internal Medicine IV, University Hospital, Trieda SNP 1, 040 66 Košice, Slovakia*

Summary: A case of hereditary nephritis with mild reduction of renal function associated with renal tubular acidosis type IV is described. The patient was admitted with life-threatening hyperkalaemia. To our knowledge, type IV renal tubular acidosis has not been reported previously in association with Alport's syndrome in an adult patient.

Correspondence: R. Tkáčová, M.D., Department of Internal Medicine I, Medical Faculty, Šafárik University, Trieda SNP 1, 040 66 Košice, Slovakia.
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Introduction

Hereditary nephritis or Alport's syndrome is a heterogeneous group of inherited abnormalities of

basement membranes which may result in renal failure, defective hearing and lens abnormalities.¹ Recently, a patient with previously diagnosed perceptive deafness presented with hyperkalaemia of 9.5 mmol/l, and was found to have mild reduction of glomerular filtration rate (GFR), and ultrastructural changes of glomerular basement membranes (GBM). Type IV renal tubular acidosis (RTA) was diagnosed by urine examination after acid loading. There are reports of this tubular dysfunction in children with nerve deafness,^{2,3} but we have found no report on type IV RTA associated with Alport's syndrome in an adult patient.

Case report

A 38 year old man of Gypsy origin was admitted to hospital in February 1992 with progressive muscular weakness, abdominal pain and vomiting for 5 days. In his family history, sudden death had occurred in two brothers and one sister, apparently as a result of a renal disease. The patient's history revealed a suggestion of Alport's syndrome in 1987 during hospitalization in a district hospital, based on the confirmation of defective hearing and microscopic haematuria; serum potassium levels were up to 6.4 mmol/l at that time.

On admission, he had a pulse rate of 60 beats/minute, and systolic blood pressure of 100 mmHg. Physical examination revealed asthenia, muscular weakness, and diffuse abdominal tenderness. His jugular venous pressure was 6 cm H₂O. There were 14 mm high T waves in V₂-V₃ leads on the electrocardiogram. Laboratory investigations showed extremely high serum potassium level (9.5 mmol/l), white blood cell count (WBC) 19.1×10^9 /l, haemoglobin 131 g/l, creatinine 155 μ mol/l, urea 11.2 mmol/l, sodium level 136 mmol/l, and phosphorus level 1.46 mmol/l. The laboratory evidence of hyperchloraemic metabolic acidosis with blood pH 7.287, base excess -8.3 mmol/l, PCO₂ 4.97 kPa, bicarbonate level 17.2 mmol/l, and serum chloride level of 113 mmol/l was present.

Haemodialysis was performed three times, each for 3 hours duration, within the first 24 hours. Other therapeutic methods were not considered in the period of life-threatening hyperkalaemia. After haemodialysis sessions serum potassium level decreased to 4.9 mmol/l, and T waves to 5 mm. No hypotensive episodes occurred during haemodialysis, and systolic blood pressure remained 100-110 mmHg also in the next period at hospital. Consequently, the patient was given treatment with frusemide at the dose of 20 mg and NaHCO₃ of 1,500 mg per day, resulting in serum potassium levels in the range of 4.4-5.3 mmol/l. The patient was stable 6 months after discharge from hospital.

Investigations

Daily urine was 1,200 ml, creatinine clearance was 0.78 ml/second. Urinalysis showed haematuria (400 million erythrocytes in Addis sediment) with no white cells, casts or proteins. Natriuresis was 93 mol/24 hours, urine potassium excretion (11-18 mmol/24 hours) were decreased, and calcium excretion (0.7 mmol/24 hours), phosphorus excretion revealed increased values (32.4 mmol/24 hours). Dietary phosphorus intake was up to 1.6 g/24 hours.

The diagnosis of type IV RTA associated with Alport's syndrome was suspected. The urinalysis dynamic tests showed the basal urine pH value 5.7, the administration of 10% calcium chloride at the dose of 57 ml led to the urine pH of 5.14, fractional excretion of bicarbonate was 0.08%. Acid loading test revealed normal excretion of phosphates (550 nmol/second/1.73 m², normal >300 nmol/second/1.73 m²) whereas ammonium excretion was inadequately low (137 nmol/second/1.73 m², normal >500 nmol/second/1.73 m²). Basal plasma renin activity (PRA) was 0.9 ng/ml/hour (normal 0.5-1.5 ng/ml/hour), aldosterone (ALD) was 25.55 pg/ml (normal 7.5-160 pg/ml), atrial natriuretic factor (ANF) was 5.74 pg/ml (normal 2-7 pg/ml). All hormonal activities were measured using radioimmunoassay analysis. After postural stimulation combined with frusemide at the dose of 80 mg the following hormone values were measured: PRA, 0.98 ng/ml/hour; ALD, 47.21 pg/ml; ANF, 2.6 pg/ml. The basal cortisol level was 750 nmol/l, 120 minutes after ACTH administration (Synacthen, CIBA) at the dose of 2 mg it increased up to 1,360 nmol/l, and thus primary hypocorticism was excluded.

Audiogram confirmed perceptive deafness affecting both ears. A computed tomography scan of adrenals demonstrated no pathological changes. On ultrasound both kidneys appeared to have normal size (left: 95 × 40 mm, right: 94 × 45 mm) with parenchyma thickness of 14 mm; no pathological echo structures were found. At light microscopic examination kidney biopsy showed nonspecific changes with pronounced interstitial fibrosis, some areas of canalicular atrophy, and arteriosclerotic changes of the arterioles with their focal fibrinoid dystrophy. Ultrastructural features of the glomeruli showed segmental mesangial proliferation, segmental sclerosis and epithelial proliferation. GBM thickness varied significantly, and some areas of extremely thin GBM were found (Figure 1).

Discussion

The diagnosis of Alport's syndrome in the present case report was based on the criteria according to

Figure 1 Marked focal thinning of the capillary basement membranes (arrows) and foot process fusion on the visceral epithelial cells. Electron microscopy (original magnification $\times 14,000$).

Genova *et al.*⁴: (1) perceptive deafness; (2) ultrastructural abnormalities of GBM; (3) positive family history. This syndrome represents a heterogeneous group of inherited abnormalities of basement membranes that may result in progressive renal failure.¹ It is suggested to be a result of mutations in an X-chromosome-encoded basement membrane collagen chain, and its phenotypic heterogeneity probably arises from allelic mutations at a single genetic locus.⁵ Thinning and splitting of the GBM are assumed to be characteristic, but nonspecific, ultrastructural alterations in Alport's syndrome.⁶ Extremely thin GBM areas were also found in our patient, and according to Rumpelt this is the basic lesion representing a persistent embryonal status of the lamina densa in Alport's syndrome.⁶ Recently, some variants of Alport's syndrome were described including oesophageal, tracheobronchial and genital leiomyomatosis, and

hereditary macrothrombocytopenia.^{7,8} However, no signs of these pathological conditions were present in our case.

Type IV RTA is a syndrome of tubular dysfunction manifested clinically by persistent hyperkalaemia and metabolic acidosis that occurs usually in patients with mild to moderate chronic renal failure.⁹ Hyperkalaemia might contribute to the acidosis by limiting urinary buffer, but the primary defect is the reduced mineralocorticoid effect on hydrogen ion secretion.¹⁰ Apart from primary hypocorticism, pseudohypoaldosteronism resulting in hyperchloraemic metabolic acidosis with hyperkalaemia may occur in different pathological conditions, such as diabetic nephropathy,¹¹ idiopathic interstitial nephritis,¹² chronic pyelonephritis,¹³ and in obstructive uropathy.¹⁴ A case of type I (hypokalaemic) RTA was described in a male child with neural deafness in whom addition of NaHCO_3 and potassium to diet improved physical condition.² Recently, discordant manifestations of RTA syndrome combined with nerve deafness in dizygotic twins were reported.³ However, for the first time to our knowledge, a case of Alport's syndrome in an adult man has been reported in association with type IV RTA resulting in hyperkalaemia. Our patient did not increase NH_4 secretion after acid loading adequately, whereas his PRA, ALD and ANF levels were within the physiological ranges.

This case illustrates that, though rare, RTA type IV has to be considered in Alport's syndrome with mild reduction of renal functions. Once considered, all available diagnostic means have to be used in order to reach a correct diagnosis and treatment and thus to prevent life-threatening hyperkalaemia.

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