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심한 간질신장염을 보인 당원축적병 type 1

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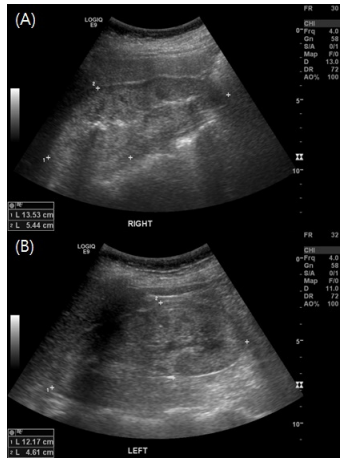
서론: 당원축적병 (Glycogen storage disease) type 1은 매우 드문 유전병으로 글리코겐 분해와 합성에 관여하는 glucose-6-phosphatase 효소 결핍에 의해서 발생한다. 간, 신장에 글리코겐이 과도하게 축적되며 신장에서는 주로 국소분절성 사구체 경화증과 동반된 세뇨관 위축을 일으키는 것으로 알려져 있다. **증례:** 16세 여자환자는 생후 5개월경부터 구토, 간비대가 발생하여 소아과에서 시행한 검사에서 당원축적병 type Ia형 진단되었다. 옥수수 전분을 이용한 식이요법으로 치료하며 정상발달을 보였으나, 최근 2년간 병원에 방문하지 않았다. 내원 당시 환자는 전신 쇠약감을 호소하였다. 한약이나 건강식품은 복용하지 않았다. 혈액검사에서 Hb 6.0 g/dL, Hct 18.0 %, sodium 138 mmol/L, potassium 2.4 mmol/L, chloride 98 mmol/L, total CO_2 7 mmol/L, BUN 81.0 mg/dL, creatinine 6.41 mg/dL, eGFR 8.85 ml/min/1.73m², uric acid 10.3 mg/dL, lactic acid 2.3 mmol/L, 24hrs urine total protein 314.2 mg/day 이 확인되었다. 복부 초음파에서 양측 신장의 크기 증가되었다(figure1). 심한 대사성 산증을 보이므로 혈액투석을 시작하였다. 신부전 원인을 알기 위해 신 조직검사를 시행하였다. 신 조직검사에서 신장세뇨관과 간질에 심한 염증세포 침윤이 있었으며 사구체 34개 중 1개에서 분절성 사구체 경화증이 있었다. **결론:** 당원축적병 type 1a에서 심한 간질성 신장염을 보인 환자를 경험하였기에 이를 보고하는 바이다.



■ Sun-304 ■

A Case of Unrecognized Diabetic Ketoacidosis aggravated by Sodium-Glucose Cotransporter 2 Inhibitor.

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Background: Sodium-glucose cotransporter 2 inhibitor (SGLT2-I) is a recently developed therapeutic agent for type 2 diabetes mellitus. SGLT2 inhibitors control blood glucose level via inhibition of renal glucose reabsorption. Clinical trials have shown that, in patients with diabetes, SGLT2-inhibitors significantly increase their survival and reduce blood pressure and body weight beside lowering blood glucose. However, SGLT2-I has some side effects such as genital and urinary tract infections, dehydration, and exacerbation of hyperkalemia. In a rare case, they may induce euglycemic ketoacidosis. Recently, in US, FDA warned development of diabetic ketoacidosis (DKA) in patients taking SGLT2 inhibitor. We present a case of DKA in a patient taking SGLT2-I, who was successfully managed by using emergency hemodialysis and insulin. **Case:** On March 1st 2018, a 36-year-old male visited emergency room with symptoms of nausea and vomiting. The patient was newly diagnosed with DM from HbA1C 13.1%. Three days ago, metformin and empagliflozin were prescribed. The next day after starting DM medication, his nausea started. On emergency room, blood pressure was 144/95mmHg, pulse was 102 beats/min, respiration rate was 28/min, and body temperature was 36.9°C. The blood chemistry showed as the following ㅡ Hb19.1, BUN 16.0, Cr 1.53, Na 138.7, K 4.59, total CO2 7, pH 7.15, pCO2 19.2, pO2 44 BE -22.0, HCO3 6.7. The patient was diagnosed with metabolic acidosis accompanied by acute kidney insufficiency (AKI). Saline hydration and bicarbonate were infused. Because patient's condition was aggravated, hemodialysis was done. On the admission day, 1 IU/hr of insulin was infused. Alongside, 50% glucose was infused for hypoglycemia. Also, hourly BST and frequent pH and bicarbonate level were monitored. On the 3rd day of admission, continuous infusion of insulin and bicarbonate was stopped. On the 4th day of admission, the patient was transferred to general ward. From then on, his blood glucose level was controlled with multiple subcutaneous insulin injections. On the 8th day of admission, he was discharged with metformin and long acting insulin.