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OLGU SUNUMU/CASE REPORT

An ignored cause of chronic kidney disease in children: type 2 cardiorenal syndrome

Çocuklarda kronik böbrek hastalığının ihmal edilen bir nedeni: tip 2 kardiyorenal sendrom

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Abstract

Cardiorenal syndrome is a disorder of the heart and kidneys in which acute or chronic dysfunction in one organ may induce acute or chronic dysfunction in the other organ. It is well known that the main cause of mortality among patients with end-stage renal disease is due to cardiovascular events and a common complication in patients in acute heart failure is a decrease in renal function. However, when there are no signs and/or symptoms of chronic cardiovascular disease, cardiovascular causes in the etiology of chronic kidney disease is not the first differential considered. We present an 11 year-old girl patient, diagnosed with type 2-chronic cardiorenal syndrome who had previously been followed in another center with the diagnosis of chronic kidney disease for six months and referred to our hospital for kidney biopsy. We present this case to increase awareness of pediatrician and nephrologist about this syndrome.

Key words: Children, chronic kidney disease, cardiorenal syndrome

Öz

Kardiyorenal sendrom hem kalp hem de böbreklerin etkilendiği bir hastalıktır ve bu organlardan birinde olan akut ya da kronik fonksiyon bozukluğunun diğer organda akut yada kronik fonksiyon bozukluğuna yol açması olarak tanımlanmaktadır. Son dönem böbrek yetmezliği olan hastaların en önemli ölüm nedeninin kardiyovasküler olaylar olduğu ve akut kalp yetmezliği olan hastalarda böbrek fonksiyonlarında azalmanın yaygın bir komplikasyon olduğu çok iyi bilinmektedir. Buna karşın, kronik kalp hastalığı olan bir hastada kardiyovasküler hastalık semptom ve bulguları belirgin değilse, kronik böbrek hastalığı etiyolojisinde kardiyovasküler hastalıklar hemen akla gelmemektedir. Bundan dolayı biz bu raporda hem çocuk hem de çocuk nefroloji hekimlerinin bu sendroma farkındalıklarını artırmak amacıyla daha önce bir dış merkezde 6 ay boyunca kronik böbrek hastalığı tanısı ile izlenen; böbrek biyopsisi yapılmak üzere hastanemize sevk edilen ve tip 2 kardiyorenal sendrom tanısı koyulan 11 yaşında kız hasta sunulmuştur.

Anahtar kelimeler: Çocuk, kronik böbrek hastalığı, kardiyorenal sendrom

INTRODUCTION

Cardiorenal syndrome (CRS) is defined as a complex pathophysiological disorder of the heart and the kidneys in which acute or chronic dysfunction in one organ may induce acute or chronic dysfunction in the other organ[1-2]. This syndrome has been classified into five groups on the basis of the primary organ dysfunction (heart or kidney) and on whether the organ dysfunction is acute or chronic[2].

Cardiovascular disease (CVD) is a major problem in patients with chronic kidney disease (CKD). In addition, renal dysfunction is also prevalent in patients with cardiac disease and is a significant

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predictor of prognosis in cardiac patients. Therefore, early identification of cardiac dysfunction in patients with CKD and renal dysfunction in patients with acute decompensated heart failure may help guide treatment decisions. However, we argue that the diagnosis of CKD is difficult in nonsymptomatic chronic CVD patients, as chronic CVD may not be the first differential considered in the etiology of CKD.

CASE REPORT

An 11 year-old girl patient, diagnosed with type 2 CRS, had previously been followed in another center with the diagnosis of CKD for six months referred to our clinic. Her past medical history revealed that the patient was healthy up until six months prior to her presentation, with the exception of a viral infection, followed by an elevation of her BUN and creatinine, between 50-100 mg/dl and 0.9-1.5 mg/dl, respectively. An extensive work-up, including voiding cystourethrography, renal cortical scintigraphy, renal ultrasonography, viral markers, complements levels, antinuclear antibody (ANA), and anti-double stranded-DNA(Anti-ds-DNA) levels were all normal. The patient had only mild proteinuria and no hematuria observed on urine analysis. The patient was then referred to our hospital for a kidney biopsy. The family gave a history including a intermittent cough of six months duration and increased fatigue. There was significant evidence of cardiomegaly and pulmonary congestion on her telecardiogram (Figure 1). Echocardiography showed severe left ventricular systolic dysfunction and an ejection fraction of 30%. The patient was diagnosed with dilated cardiomyopathy.

The laboratory results were as follows; calcium level 10.1 mg/dL (normal range, 8.5 to 10.5mg/dL), serum phosphorus level 5.4 mg/dL (normal range, 2.0 to 5.0mg/dL), alkaline phosphatase 166 UI/l (normal range, 37 to 147 IU/lt), and parathyroid hormone level 832 pg/mL. Her detailed history revealed that she was born full term after a twin pregnancy and was the child of related (third degree of kinship) healthy parents. Her twin brother was healthy. In addition to her twin brother, she had four siblings. All of her siblings were boys. One of them died at six years old due to an unknown etiology. Upon physical examination, height and weight were between the 75-90 and 25-50 percentile, respectively. Her pulse rate was 90/min, respiratory rate 22/min, body temperature 36.5°C, blood pressure 110/70 mmHg. There was no audible murmur or pathologic heart sounds on auscultation of heart and no audible crackles on auscultation of the lungs. There was only mild hepatomegaly.

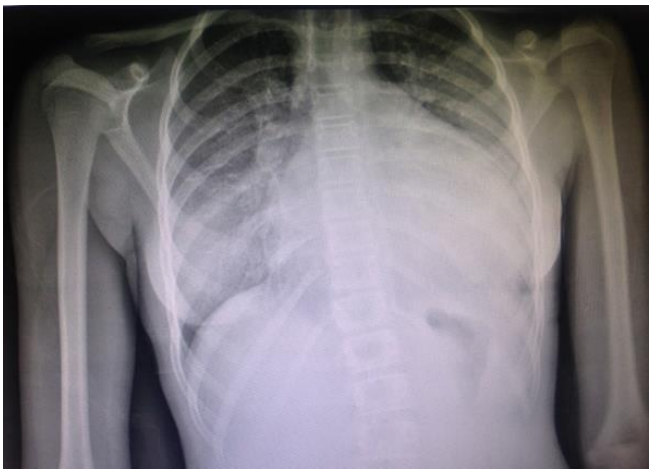

Figure 1. Telecardiography of patient; there is prominent cardiomegaly and pulmonary congestion

DISCUSSION

Interactions between cardiac and renal function have been explained by haemodynamic factors such as hypoperfusion of the kidneys and decreased venous return leading to venous congestion[1]. These factors lead to a decrease in renal functions in acute or chronic decompensated heart failure. In addition, longstanding heart failure may lead to significant

renal fibrosis and irreversible renal damage despite hemodynamic improvement[3]. Thus, preventing these factors becomes an important long-term goal in the treatment of CRS4. The association between heart failure and renal insufficiency has been demonstrated in various studies[5]. It is well known that CVD is the most common leading cause of morbidity and mortality in patients with renal dysfunction[6]. Therefore, all patients with the diagnosis of CKD are followed by physicians for CVD; subsequently, the early identification of CVD is possible in these patients[7]. In addition, according to previous studies, renal insufficiency is present in more than one-third of patients with acute decompensated heart failure[8]. Further, it is well known that a decrease in renal function in acute decompensated heart failure is an independent predictor of mortality[9]. Therefore, physicians also evaluate these types of patients for the risk of decreasing renal function in their outpatient clinic. On the other hand, we argue that there is problem in the evaluation of a patient with the diagnosis of CKD secondary to chronic abnormalities in heart function. This type of CRS is known as Type 2 CRS, chronic cardiorenal syndrome. Often in the evaluation of Type 2 CRS patients, chronic CVD in the etiology of CKD is not the first differential considered. This cognitive error leads to a delay in the diagnosis and treatment of these patients. A delay in the diagnosis also leads to progression of the disease. With an increased prevalence of chronic cardiorenal syndrome due to improved survival of patients with heart failure[10], it is prudent to consider the diagnosis of CVD when evaluating the patients with chronic renal injury or dysfunction. This is particularly important for optimal care of patients without a diagnosis of pre-existing CVD as in our case and it provides a more prompt diagnosis and treatment for type 2 CRS patients. Cooperation between pediatric cardiologists and pediatric nephrologists is crucial to better understand this syndrome.

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