

Congenital Nephrotic Syndrome of The Finnish Type with Crescent Formation: A Challenging Case and Ultrasonographic Findings

Al-Mosawi AJ*

*Department of Pediatrics and Pediatric Psychiatry, Children Teaching Hospital of Baghdad Medical City, Iraq

1. Abstract

Background: Congenital nephrotic syndrome of the Finnish type has been probably observed in Iraqi children, but it was not reported or documented in the scientific literature. The aim of this paper is to report the rare occurrence of congenital nephrotic syndrome of the Finnish type in association with crescents formation. Interesting ultrasonographic findings are also reported, and an evidence-based therapeutic recommendation for Iraqi patients is described.

Patients and methods: A four-month male infant with congenital nephrotic syndrome was referred with a request for therapeutic recommendation. Ultrasound study was performed. The ultrasound specialist thought that the ultrasound diagnosis could be medullary sponge kidney because of the presence of hyperechoic medulla. The ultrasound finding was considered, but the ultrasound specialist diagnosis was not taken as it was not correlated with the clinical diagnosis of nephrotic syndrome.

Results: The ultrasound finding was considered, but the ultrasound specialist diagnosis was not taken as it was not correlated with the clinical diagnosis of nephrotic syndrome. Renal biopsy showed the classic diagnostic histopathological findings congenital nephrotic syndrome of the Finnish type. In addition, biopsy showed the rare occurrence of

crescents formation in the congenital nephrotic syndrome of the Finnish type.

Conclusion: The rare occurrence of congenital nephrotic syndrome of the Finnish type in association with crescents formation is reported and supplemented with interesting ultrasonographic findings.

2. Keywords: Congenital Nephrotic Syndrome, Finnish Type, Crescents Formation

3. Introduction

Congenital nephrotic syndrome was first reported by Gautier and Miville in 1942, and is typically presented with heavy proteinuria ($>200\text{mg}/\text{mmol}$ creatinine), hypoalbuminemia, and edema, occurring in the first three months of life. It is mostly genetic condition, and the Finnish-type is also called "Infantile microcystic disease" is probably the single most important type [1, 2].

Congenital nephrotic syndrome of the Finnish type has been probably observed in Iraqi children, but it was not reported or documented in the scientific literature [3, 4]. The aim of this paper is to report the rare occurrence of congenital nephrotic syndrome of the Finnish type in association with crescents

***Corresponding author:** Al-Mosawi AJ, Department of Pediatrics and Pediatric Psychiatry, Children Teaching Hospital of Baghdad Medical City, Iraq, E-mail: almosawiAJ@yahoo.com

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formation. Interesting ultrasonographic findings are also reported, and an evidence-based therapeutic recommendation for Iraqi patients is described.

4. Patients and methods

A four-month male infant with generalized edema and heavy proteinuria appearing before the age of three months was referred with a request for therapeutic recommendation. The parents were consanguineous, but family history was negative for a similar disorder. No information was available about the size of placenta at birth.

Renal ultrasound showed that the right kidney was 7.8 X 3.5 cm in size, and had smooth outline (Figure 1), renal parenchyma thickness was reduced. The right kidney had echogenic pyramids, and mildly increased cortical echogenicity. The pelvi-calyceal system was normal.

Renal ultrasound of the left kidney showed was 8.6 X 4.3 cm in size, and had smooth outline (Figure 2), renal parenchyma echo-texture was similar to that of the right kidney. The pelvi-calyceal system was normal.



Figure 1: Renal ultrasound showed that the right kidney was 7.8 X 3.5 cm in size and had smooth outline, renal parenchyma thickness was reduced. The right kidney had echogenic pyramids, and mildly increased cortical echogenicity. The pelvi-calyceal system was normal.



Figure 2: Renal ultrasound of the left kidney showed was 8.6 X 4.3 cm in size, and had smooth outline, renal parenchyma echo-texture was similar to that of the right kidney.

Abdominal ultrasound showed ascites and abdominal organs (liver, spleen, pancreas, gall bladder) were normal in size and texture.

The ultrasound specialist thought that the ultrasound diagnosis could be medullary sponge kidney because of the presence of hyperechoic medulla. The ultrasound finding was considered, but the ultrasound specialist diagnosis was not taken as it was not correlated with the clinical diagnosis of nephrotic syndrome.

5. Results

The ultrasound finding was considered, but the ultrasound specialist diagnosis was not taken as it was not correlated with the clinical diagnosis of nephrotic syndrome. Renal biopsy was performed and 26 glomeruli were examined with light microscope and showed the classic diagnostic histopathological findings congenital nephrotic syndrome of the Finnish type including tubular microcystic changes and diffuse mesangial proliferation. There were also tubular hyaline casts, and thickened small blood vessel walls. In addition, biopsy showed the rare occurrence of crescents formation in the congenital nephrotic syndrome of the Finnish type. The basement membrane thickness was normal.

6. Discussion

The occurrence of congenital nephrotic syndrome of the Finnish type in association with crescents formation has been very rarely reported in the literature Ghosh et al [5].

Ultrasonographic findings of congenital nephrotic syndrome of Finnish type have been infrequently described in the literature Graif et al, Perale et al, Alkrinawi et al, Lanning et al, Bratton et al, Northrup et al [6-11].

Lanning et al (1989) described the ultrasonic findings of seven infants with congenital nephrotic syndrome of the Finnish type who was on active conservative treatment. The kidneys were enlarged and the cortex more echogenic than the liver and spleen parenchyma. The pyramids were small with hazy borders resulting in an indistinct or lost corticomedullary border. Follow-up showed increased the renal cortex echogenity in four infants and the pyramids became invisible [9].

In this case, the ultrasound specialist thought that the ultrasound diagnosis could be medullary sponge kidney because of the presence of hyperechoic medulla, but we took the ultrasound findings and excluded the ultrasound specialist's diagnosis for being not correlated with the clinical diagnosis of nephrotic syndrome. However, Bratton et al reported a case of congenital nephrotic syndrome, Finnish type, which was initially misdiagnosed as infantile polycystic kidney disease based on ultrasonographic findings [10].

Management of congenital nephrotic syndrome has been considered challenging even in developed countries where bilateral nephrectomies followed by dialysis and transplantation represents the standard therapeutic recommendation and are practiced in most centres. In this country, these practices are not applicable, and evidenced based conservative treatment have to be used [12, 13].

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