Nephropathy in a child suffering from tetralogy of Fallot

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Received - 21 March 2020 Initial Review - 23 April 2020 Accepted - 17 May 2020

ABSTRACT

Cyanotic nephropathy is a type of renal impairment, occurring due to chronic exposure to hypoxia, seen in unoperated cases of cyanotic congenital heart diseases (CCHDs). Due to a paucity of the literature and lack of general awareness of this condition, many times, the occurrence of this nephropathy is left untreated, leading to chronic kidney disease (CKD). The mortality rate is 51% in children who suffer from CCHD with CKD. Here, we report a case of an 11-year-old male child with tetralogy of Fallot (unoperated), requiring repeated admissions for breathlessness. An incidental finding of proteinuria on multiple admission was further investigated. The patient had hemoconcentration with deranged renal function tests, [hypoalbuminemia (2.2 gm/dl), serum creatinine 3.4 mg/dl, eGfR 16.28, and urine protein:creatinine ratio 3.95]. USG KUB done reported a bilateral medicorenal disease with a complete loss of corticomedullary differentiation. The patient already had Stage IV CKD at the time of presentation and expired 8 months after diagnosis.

Key words: *Cardiorenal syndrome, Congenital heart disease, Cyanotic nephropathy, Tetralogy of Fallot*

Cyanotic congenital heart disease (CCHD) with renal impairment is known as cyanotic nephropathy (CN), which is a result of chronic exposure to hypoxia [1]. This is an example of cardiorenal syndrome (CRS) type 2, which if left untreated, may lead to progressive chronic kidney disease (CKD), the mortality being 51% in children suffering from both [1].The extent of glomerular damage is related to the duration of cyanosis and elevated values of hematocrit [2].

Here, we describe the case of an 11-year-old male child with tetralogy of Fallot (ToF) (not operated), with poor follow-up leading to incidental findings of nephropathy. This case has been reported for the need of a multispecialty approach for early detection and management of CN.

CASE REPORT

An 11-year-old male child presented at our center with complaints of breathlessness and fever for 1 day. The patient was diagnosed with TOF at 9 months of age and was on palliative treatment due to the refusal of corrective surgery. The patient was on long-term treatment with propranolol, torsemide, and enalapril.

On admission, the patient's vitals were – heart rate 110/min, respiratory rate 38/min, blood pressure 110/60, and saturation 82% on FiO_2 of 28%. Physical examination revealed respiratory distress with peripheral cyanosis and clubbing of his fingers and toes. There was no edema. His chest X-ray showed the classical "boot-shaped heart" with an upturned cardiac apex (Fig. 1). Echocardiography revealed ToF with an atretic pulmonary valve and severely hypoplastic pulmonary artery but with normal biventricular function.

The patient's routine investigations revealed hemoconcentration (hemoglobin 155 g/L and packed cell volume 46.9), with deranged renal function tests (blood urea nitrogen 18.92 mmol/L, serum creatinine 300.56 micromole/L, and eGFR 16.28), proteinuria (urine albumin on dipsticks examination 4+ and urine protein:creatinine ratio 3.95), and hypoalbuminemia (22 g/L). He had normal serum cholesterol (4.97 mmol/L) and his electrolytes were suggestive of mild hyponatremia and hyperkalemia (serum sodium 134 mmol/l, serum potassium 5.6 mmol/l, and serum chloride 109 mmol/l). Ultrasonography of the kidney, ureter and bladder (KUB) reported small-sized kidneys (right kidney 6 × 3.1 cm, left kidney 6.6 × 3.2 cm) with complete loss of corticomedullary differentiation, favoring a diagnosis of chronic nephropathy secondary to hypoxic damage due to CCHD.

Due to the CKD Stage 4, enalapril was discontinued and was advised supportive CKD management (since the dyselectrolytemia and other solute load were easily controllable on oral medication). The parents were also counseled for regular follow-up and a probable need for long-term hemodialysis in the future. The patient was stable on discharge but expired 8 months later. His autopsy report showed an enlarged heart with left ventricular hypertrophy with 80% obliteration of the lumen of the left anterior descending coronary artery.

DISCUSSION

Our patient presented late, having already developed CN. Early detection and management of proteinuria would have led to a slower progression of renal damage. Due to financial constraints

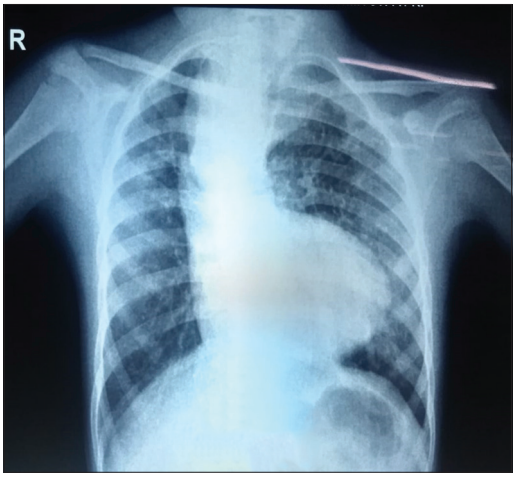

Figure 1: Chest X-ray showing "Boot-shaped" heart

and educational ignorance, parents did not maintain a regular follow-up even while the child was on multiple drugs including enalapril for palliative care of ToF. Early changes of nephropathy such as proteinuria and the early rise of creatinine were missed. On presentation to our center, he was already in Stage IV chronic kidney injury.

In CN, glomerulomegaly is an important pathogenesis, leading to the decline of kidney function [3,4]. CN is seen in 30–50% of patients with CCHD affecting both tubular and glomerular functions . The resulting effect is seen as proteinuria and azotemia [3,6]. Hypoxia, polycythemia, and hyperdynamic circulation seem to be important factors deciding the rate of progression of nephropathy [7,8]. Studies have shown that phlebotomy helped to decrease proteinuria along with reducing the hematocrit [9,10]. This may be due to a reduced blood viscosity which seems to exert beneficial effects on hypoxia and glomerular permeability [11,8].

Untreated cardiac malformations in patients with CCHD have a high likelihood of progression to glomerular damage [12]. The risk of developing glomerular lesions rises sharply during the second decade of life if the cyanosis remains unchanged for more than 10 years [13,14]. A study done by Hongsawong *et al*. involving 94 patients, aged 1 month–15 years with CCHD, found the prevalence of CN based on significant proteinuria or albuminuria to be 92.55% and 58.51%, respectively. They concluded that CN in CCHD can be detected as early as the 1st decade of life, by doing annual screening using, early morning urine samples for urinalysis, urine protein, and albumin [15].

CONCLUSION

In our patient, the unoperated ToF led to CN. A multispecialty approach involving, pediatric cardiologists, nephrologists, and intensivists, is necessary for the management of CRS in CCHD patients, to improve the long-term outcomes in these high-risk patients. Early corrective surgery is recommended to halt complications due to CRS. Children with CCHD should undergo regular screening for the detection of renal dysfunction.

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Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Naik VS, Phadke VD, Kanhere SV. Nephropathy in a child suffering from tetralogy of Fallot. Indian J Case Reports. 2020;6(6): 312-313.

Doi: 10.32677/IJCR.2020.v06.i06.009