Nephrocalcinosis in a preterm infant

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Abstract Bilateral medullary echogenic foci are not uncommonly seen in renal ultrasounds of sick, preterm neonates. Often, the likely diagnosis is nephrocalcinosis. We present one such case. The long term outcome in this population is unknown, but is generally thought to be essentially good.

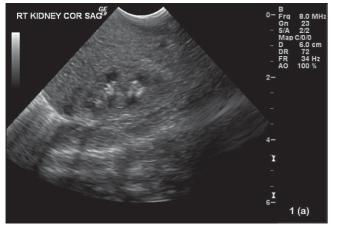




Fig. 1: bilateral medullary echodenisities (a) right kidney (b) left kidney.

Introduction

Nephrocalcinosis refers to the deposition of calcium within the wall and lumen of the distal collecting tubules and is detectable by ultrasound in 20–64% of preterm infants. Multiple factors have been implicated in the aetiology of this phenomenon. However, there has been limited study of the long term effects. Ultrasound remains the first line imaging modality for assessment of the neonatal kidney and is the preferred modality for assessing calcification in preterm infants^{1,2}. Understanding the appearance and differential diagnosis of nephrocalcinosis on ultrasound of the preterm kidney may aid in management.

Case history

The case presented is that of a female infant, born at 26 weeks gestation, birth weight 963 g. She required respiratory support with continuous positive airways pressure (CPAP) and developed moderate bilateral intraventricular haemorrhages. She had a patent ductus arterious, persistent metabolic acidosis and significant hyponatraemia. She was treated with Indomethacin and fluid restriction in week one, then sodium and bicarbonate supplements. At three weeks of age, she developed sepsis with severe hyponatraemia, persistent metabolic acidosis, with significant urinary sodium and bicarbonate loss.

Renal ultrasound demonstrated kidneys of normal size for gestation. Multiple echogenic foci without acoustic shadowing were seen in the region of the tips of the medullary pyramids bilaterally (Fig. 1). Corticomedullary differentiation was otherwise preserved. No pelvicalyceal dilatation was identified on either side. The bladder outline was normal. The echogenic foci were unchanged on repeat examination two weeks later.

Discussion

The ultrasound appearance of the kidneys in neonates differs significantly from that in older children. The renal cortex has echogenicity equal to or greater than that of the liver and spleen, whereas in older children and adults, the cortex is relatively hypoechoic. The echogenicity of neonatal renal cortex is due to the relative concentration, as well as the increased cellular volume, of glomeruli. The medullary pyramids appear prominent and hypoechoic because of a relatively lower cortical volume³.

Increased medullary echogenicity in the preterm infant is a non-specific finding and may be due to a number of problems such as nephrocalcinosis, the sloughed papillae of papillary necrosis, vascular congestion, fungal infection or early transient protein cast deposition (Tamm-Horschfall proteinuria)⁴⁻⁶.

The infant described in this case had non-specific findings of tubular dysfunction and haematuria. When correlated with the clinical history, identification of increased echogenicity in the medulla, unchanging over two weeks, although not a definitive diagnosis, suggests nephrocalcinosis as the likely diagnosis of the ultrasound findings. Serial ultrasounds would have been helpful in establishing this more definitively.

Infants affected by nephrocalcinosis tend to be sicker infants, who are more likely to have periods of acidosis, or to have received treatments such as diuretics and steroids⁴. The ultrasound appearance of increased medullary echogenicity tends to be symmetrically bilateral, and may range from hyperechogenicity at the tip of the pyramid or hyperechoic rim around the pyramid to intense hyperechogenicity filling the pyramid entirely⁵.

Long-term effects remain unknown, although many



studies have suggested the majority have resolved by mid childhood. A small study of 14 very low birth weight infants followed to seven years reported no measurable differences in renal function compared with a control group. Others have reported renal calculi in a few affected children. In one recent study of 40 preterm infants nephrocalcinosis was associated with poorer glomerular and tubular function at seven years^{12,4,5,7}.

Renal papillary necrosis (RPN) is the major differential diagnosis in this clinical situation. RPN can present with haematuria, features of urinary infection, renal obstruction or colic. Occasionally, it may present as acute renal failure and lead to chronic renal failure. Increased medullary echogenicity may be patchy or diffuse, unilateral or bilateral, and tends to be more extensive as severity increases⁶. RPN can be distinguished from nephrocalcinosis on ultrasound with time as RPN shows distortion of the papillae and sloughing within two weeks, whereas nephrocalcinosis tends to remain unchanged. In adults, it has been related to the use of nonsteroidal anti-inflammatory drugs, as well as diabetes, dehydration/hypotension, renal vein thrombosis, acidosis and respiratory distress. There is limited information regarding the neonatal population.

Conclusion

Nephrocalcinosis is common in the preterm population, particularly those with significant or prolonged illness. Ultrasound findings should be correlated with clinical history. Despite a degree of reassurance from the literature that these children are unlikely to suffer adverse long term consequences, further long term follow up studies are required before this can be conclusively determined. The lack of clarity from the existing literature regarding long term prognosis may justify long term follow up of these children.

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