CASE REPORT

White-blue pyelocalyceal cyst with hydrotic glomerulonephritis

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SUMMARY

A 5-month-old male infant presented with a 15 day history of distension of abdomen. On clinical examination, a soft lump was palpable in the left lumbar region. Radiological findings suggested an enlarged non-functional left kidney with ureteropelvic adhesive obstruction. The left renal mass was excised and submitted for histopathological examination. The excised renal mass was cystic with its wall partly white and partly blue. Gross and histopathological findings were diagnostic of a white-blue pyelocalyceal cyst with hydrotic glomerulonephritis. This entity needs to be differentiated from a large number of other cystic diseases of the kidney. Intrauterine screening and diagnosis may be significant for a possible early intrauterine uro-laparoscopic recanalisation of the pyeloureteral obstruction to save the affected kidney.

BACKGROUND

Pyelocalyceal cyst is a rare congenital developmental anomaly due to obstructive adhesion at the pyeloureteric junction, which irreversibly damages and defuncts the affected kidney. Only one case of white-blue pyelocalyceal cyst of the kidney has been recently reported in the literature.¹ We report diagnostic findings with differential diagnosis in another case of a left-sided white-blue pyelocalyceal cyst in a 5-month-old Indian infant.

CASE PRESENTATION

A 5-month-old male child presented with a 15-day history of progressive distension of the abdomen. The infant's birth was full-term, spontaneous, vaginal, home delivery with unremarkable perinatal and birth history. There was no history of fever, pain, urinary-tract infection, haematuria or any systemic illness or infection. There was no family history of cystic renal disease or of any other congenital anomaly. On clinical examination, the infant's abdomen was found to be soft, non-tender, with a lump in the left lumbar region. Ultrasonography of the abdomen showed enlarged left kidney measuring 82×44 mm in size, having cystic appearance, with loss of corticomedullary differentiation. The right kidney was observed to be normal, measuring 52×26 mm in size. Renal scan findings showed an enlarged left kidney with thinned out cortex. Uptake of the tracer was reduced. The corticocalyceal transit was delayed and there was no appreciable excretion of tracer till 2 h, and even after forced diuresis. Renographic curve of the left kidney showed loss of cortical and excretory phases. The right kidney was normal in

size and shape, cortex showed normal uniform uptake of the tracer, corticocalyceal transit was normal and there was prolonged excretion of tracer from the right kidney. Renographic curve of right kidney showed normal cortical and prolonged excretory phases. Radiological findings suggested evidence of adhesive obstruction in the left ureter with cystic left kidney and severely compromised cortical function. The non-functional left renal mass was excised by the surgeon by transperitoneal laparoscopic radical nephrectomy. The renal artery and vein were separately dissected, clipped and divided and the ureter was transected before excising the renal mass. The excised specimen was submitted for the final diagnosis.

On gross examination, the excised renal mass was cystic with surface lobulations. On the surface, a white lining wall of pyelocalyceal cyst appeared joined with the blue wall of the cystic kidney (figure 1). The cut surface showed confluent cyst with five lobulations with the largest measuring 10 cm in diameter, and its wall was contributed by the white lining of the pelvic part of the cyst, and the blue lining of calyceal cysts occupying the kidney with thinned out compressed renal cortical tissue (figure 2). The cyst wall was parchment-like with a thickness of about 1–2 mm. The pyelocalyceal cyst lumen was filled with clear watery fluid. The ureter was atretic and its lumen was obliterated with adhesion at the pyeloureteric junction.

On microscopic examination, H&E-stained sections from the pelvic (white) part of the cyst wall comprised oedematous fibromuscular tissue lined on the inner side by attenuated transitional epithelium (figure 3). The calyceal (blue) part of the cyst

Figure 1 Pyelocalyceal cyst showing prominent surface lobulations with white pelvic (arrow) and blue calyceal (double arrow) parts.

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Figure 2 Cut surface showing confluent cyst with lobulations. Cyst wall is contributed by white lining wall of the pelvic cysts (arrow), and blue lining of calyceal cysts (double arrow) occupying the kidney.

wall comprised a thin strip of compressed renal tissue showing cystically dilated tubules, ballooned glomerular spaces with atrophy of the glomerular tuft and sclerosis of glomerular capillary walls (figures 4 and 5). Proximal convoluted tubules were dilated and showed cloudy swelling of the lining epithelium (figure 5). Interstitium showed mononuclear inflammatory infiltrate.

INVESTIGATIONS

Routine urine and haematological findings were within normal limits. Serum urea was 31 mg/dL and serum creatinine was 0.91 mg/dL. The patient's liver function tests were normal.

DIFFERENTIAL DIAGNOSIS

Findings in this case were diagnostic of pyelocalyceal cyst of the left kidney with ureteropelvic adhesive obstruction. Differential diagnosis included solitary renal cyst, pyelogenic cysts, pyelocalyceal diverticulum, renal sinus or parapelvic cyst, polycystic kidney disease, multicystic renal dysplasia, megacalycosis, nephronophthisis, hydronephrotic kidney, benign cystic lymphangioma, distinguishable by meticulous clinical, gross and histopathological examination findings.

Figure 3 Pelvic cyst wall (white) comprising oedematous fibromuscular tissue (double arrow) lined on the inner side by attenuated transitional epithelium (arrow; H&E, $\times 500$).

Figure 4 The calyceal cyst wall (blue) showed thin strip of compressed renal tissue having cystically dilated tubules (double arrow), ballooned glomerular spaces with hydrotic glomerulonephritis and glomerular sclerosis (arrow; H&E, $\times 120$).

OUTCOME AND FOLLOW-UP

Postoperative recovery of the patient was uneventful. The patient had been well during the past 2-month follow-up.

DISCUSSION

Histopathological findings, in the present case, were diagnostic of a white-blue, pyelocalyceal cyst with hydrotic glomerulonephritis with ureteropelvic adhesive obstruction. A similar case was earlier reported in the literature.¹ The kidneys in infants and children are known to present with various forms of single or multiple cysts, arising from the renal tubules, calyces and pelvis due to congenital or acquired obstructions at various levels. However, the significance of the pathology and pathogenesis of the pyelocalyceal cyst has remained poorly understood.^{1 2} During embryonic development, the renal glomerular apparatus develops from the metanephrogenic mass (metanephrotome). The pelvic tubular drainage portion of the kidneys develops as a ureteric bud (metanephric diverticulum) from the distal end of the mesonephric duct near the cloaca. The ureteric bud elongates, expands and branches into the metanephrogenic mass to produce the drainage pathway for urine. At the eighth week, the terminal portion of the ureteric bud expands to form the renal

Figure 5 Dilated proximal convoluted tubules with cloudy swelling of the lining epithelium (arrow) and ballooned glomerular spaces with hydrotic glomerulonephritis (double arrow; H&E, $\times 500$).

pelvis, and branches to form the calyces and associated collecting tubules of the renal medulla. During this differentiation of ureteric bud into various structures, the budding points marking the ureteropelvic, pelvi-calyceal, calyceal-tubular, collecting tubulonephric junctions are left as distinctly narrow segments, liable to result in obliterating urothelial adhesions, leading to pelvic, calyceal, ductular, tubular and glomerular-nephric cysts, depending on the site(s) of obliteration.³ In the present case, adhesive ureteropelvic obstruction gave rise to confluent pyelocalyceal-tubular, white-blue cyst. The confluent cyst of the calyces and collecting tubules occupied and compressed the kidney tissue leading to congestion and cyanosis of the renal venous channels, imparting blue discolouration. Microscopic appearance of the hydrotic nephritis with glomerulosclerosis of the nephrons, in the present case, suggested patency at the pelvi-calyceal and calyceotubular and tubulonephric junctions.

Pyelocalyceal cyst is differentiable from a solitary renal cyst which is a unilocular cyst lined by flattened cuboidal epithelium and not communicating with the pyelocalyceal system.⁴ A pyelogenic cyst or a pyelocalyceal diverticulum of the kidney is a cystic cavity lined by transitional epithelium, enclosed within the renal parenchyma, and communicating with calyceal and collecting tubules via narrow openings.⁵ Patients with pyelocalyceal diverticulum usually present with haematuria, symptoms of urinary tract infection, renal colic or obstruction due to calculus formation. Pyelocalyceal diverticulum presents ultrasonographic findings of thick-walled cysts with mobile echogenic debris due to milk of calcium and often calculus formation.⁶ These findings differentiated the pyelogenic cyst or diverticulum from the presently observed pyelocalyceal cyst.

A parapelvic cyst or a renal sinus cyst of lymphatic origin, arising in proximity to the pelvis and major calyces, can cause diagnostic confusion.⁷ The parapelvic cyst does not communicate and presents separately with the collecting system, in contrast to the white-blue pyelocalyceal cyst.

Polycystic kidney disease often presents with genetic history and is characterised by non-functioning, enlarged, diffusely cystic kidneys.⁸ Numerous small cysts in the cortex and medulla in this disease give the kidney a sponge-like appearance. Histopathologically, the cysts present with saccular dilation of the collecting tubules and are lined with the uniform cuboidal epithelium rather than the transitional epithelial lining seen in the white-blue pyelocalyceal cyst.⁹ Also, in children, polycystic kidney disease is invariably bilateral, associated with cysts in the liver along with portal fibrosis and proliferating portal bile ducts, and is incompatible with life.

Another cystic disease of the kidney is multicystic renal dysplasia, which is due to the abnormality in metanephric differentiation associated with ureteral atresia and pyelocalyceal obstruction and presents with an enlarged, irregular and multicystic kidney.^{10–11} Histopathologically, flattened epithelium lining of the dysplastic cyst and the presence of islands of undifferentiated mesenchyme, often cartilage, and immature collecting ducts distinguish it from the white-blue pyelocalyceal cyst.¹²

The white-blue pyelocalyceal cyst, in the present case, differs from the megacalycosis which is characterised by non-obstructive dilation of renal calyces, with normal pelvis and hypoplastic renal medullary pyramids.¹³

Renal cyst cortical atrophy can also be seen in the nephronophthisis, but the cyst in nephronophthisis is predominantly corticomedullary and is lined by flattened or cuboidal epithelium rather than transitional epithelium of pyelocalyceal cyst. Moreover, nephronophthisis is predominantly familial (familial juvenile nephronophthisis) with autosomal recessive inheritance

and usually manifests in children and young adults rather than in infants.

The white-blue pyelocalyceal cyst must be distinguished from the hydronephrosis, which indicates retention of urine up to the kidney, irrespective of the disease causing obstruction of urinary flow, leading to dilation of orifices and renopelvi-ureteric channels, with no universally accepted value or definition.¹⁴ In this case, the renal pelvis and calyces and collecting tubules were transformed into a confluent lobulated cyst, which give rise to hydronephritically dilated cystic nephrons, with consequent hydrotic glomerulonephritis and sclerosis of the glomerular tufts.

Although many of the congenital anomalies of the fetus may be surgically correctable, a number of these may be missed in the intrauterine period due to the lack of obvious clinical symptoms. Some anomalies, like the present one, may silently damage and defunct the organ and may eventually become life threatening. The fetus having in utero obstructive uropathy may develop pyelocalyceal cyst and severe atrophy of renal parenchyma due to pressure exerted by the retained urine with consequent loss of kidney function, manifesting shortly after birth. Routine antenatal ultrasonographic screening, and early intra-uterine ureteroendoscopic recanalisation, may possibly save the affected kidney in such cases of obstructive uropathy.¹⁵

Learning points

- ▶ Adhesive obstruction at ureteropelvic junction may give rise to white-blue pyelocalyceal cyst.
- ▶ The pelvic part of the pyelocalyceal cyst is white in contrast to the calyceal part of the cyst occupying and presenting as blue kidney.
- ▶ The white-blue pyelocalyceal cyst is grossly and histopathologically distinguishable from other cystic diseases of the kidney.
- ▶ Intrauterine screening in all cases may be advisable for early diagnosis of pyelocalyceal cyst and a possible laparoscopic recanalisation of the obstruction to save the affected organ.

Competing interests None.

Patient consent Obtained.

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