References


Retroaortic Left Renal Vein with Cascade of Complications in a Neonate

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A retroaortic left renal vein (RLRV) is located between the aorta and lumbar vertebrae, and drains into the inferior vena cava (IVC) or left common iliac vein [1]. Compression of the left renal vein between the abdominal aorta and vertebrae leads to haematuria, flank pain, varicocele and abdominal pain; this is also called posterior nutcracker phenomenon [2]. Congested kidney and renal infarcts secondary to posterior nutcracker phenomenon may lead to bacterial localization and abscess formation. Aortic thrombosis is a recognized complication of infection and sepsis [3]. Computed tomography (CT), magnetic resonance imaging and ultrasonography (USG) are effective for detection of this congenital anomaly [1]. We present a neonate with RLRV with posterior nutcracker phenomenon who subsequently developed sepsis and thrombosis of abdominal aorta.

Case Report

A 14-day-old male neonate was admitted with history of lump abdomen and excessive cry for 4 days and progressively enlarging lump in left side of abdomen for 2 days. There was no history of fever, lethargy, poor feeding, vomiting, seizure or any respiratory symptom. There was no bowel or urinary complaints. Perinatal period was uneventful.

The infant was irritable with normal general physical examination and stable vitals. There was a hard, non-mobile, 4×3 cm lump present in left hypochondrium and lumbar regions. Rest of the systemic examination was normal.

Investigations revealed neutrophilia and deranged renal function (urea 177 mg/dL; creatinine 1.4 mg/dL).
Urine examination showed plenty of red blood cells, leukocytes and proteinuria. USG abdomen showed enlarged left kidney with altered echotexture. After 2 days of admission, the child developed lethargy, poor feeding and shock. Sepsis was suspected; investigations showed neutrophilia with toxic granules, positive CRP and Staphylococcus aureus in blood culture.

The infant responded to antibiotics and inotropes but had feeble bilateral femoral and distal lower limb pulses. CT abdomen (Fig. 1 and 2) showed retroaortic left renal vein (type 1) with bulky, poorly functioning left kidney. Multiple interparenchymal non-enhancing hypodense areas were seen suggestive of multiple renal abscesses with extension into lower perirenal space; thrombus was present in lower abdominal aorta. Doppler study showed monophasic flow in femoral arteries suggestive of proximal partial obstruction. Coagulation profile of the patient was normal.

The child improved on antibiotics, low molecular weight (LMW) heparin and supportive measures with gradual disappearance of lump and improved pulse volume. Follow-up CT angiography done after 18 days showed 20% luminal narrowing of distal abdominal aorta with small residual abscess in left kidney at lower pole. On follow-up after 3 months of treatment with LMW heparin, Doppler USG showed small left kidney with residual scarring of lower pole with normal blood flow in renal artery and abdominal aorta; injection heparin was stopped.

DISCUSSION

RLRV is a malformation characterized by presence of a vessel that drains the left renal blood up to the inferior vena cava crossing behind the aorta [1]. The incidence of RLRV has been reported from 0.5% to 2.3% [4,5]. RLRV is classified into four types according to their drainage site and the most common is type I [1]. Nutcracker phenomenon refers to compression of the left renal vein, most commonly between the aorta and the superior mesenteric artery, and posteriorly between aorta and vertebrae [2]. Symptoms commonly include hematuria, flank pain, varicocele and proteinuria [6]. Hematuria is the most common symptom and is due to rupture of thin-wall varices into the collecting system, secondary to elevated venous pressure [6]. This patient with RLRV type1 presented with acute onset left renal lump, excessive cry, hematuria and proteinuria suggestive of posterior nutcracker phenomenon. These focal renal bleeds and infarcts are potential sources of bacterial localization that may result in sepsicaemia, as was seen in our patient. Thrombosis of aorta in our patient was secondary to septicemia and renal abscesses extending into perirenal space of left kidney.

We conclude that RLRV can lead to a cascade of complications due to its compression, and should be suspected in an infant presenting with renal lump and hematuria.

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Bilateral Torsion of Testes with Purpura Fulminans

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Background: Purpura fulminans and bilateral perinatal testicular torsion are rare and may co-exist. Case characteristics: A 3-day-old neonate with bilateral swelling of scrotum; torsion and gangrenous changes were observed on exploration. Interventions: Left orchidectomy with preservation of right testis was done. Outcome: At 2-month follow-up, right testis showed signs of atrophy. Child developed full thickness skin lesions and died of sepsis. Message: Perinatal testicular torsion can be bilateral, and requires urgent surgical exploration.

Keywords: Perinatal testicular torsion, Protein-C deficiency, Purpura fulminans.

P erinatal testicular torsion includes intrauterine and postnatal torsion occurring within the first 30 days of life. It is sporadic and represents about 12% of all testicular torsions during infancy [1]. Bilateral perinatal testicular torsion is even rarer [2]. Protein C deficiency leads to decreased capacity to reduce thrombin generation, which leads to a hypercoagulable state [3]. We present a neonate who developed bilateral testicular torsion and was diagnosed to be having protein C deficiency.

CASE REPORT

A late preterm neonate with severe intrauterine growth restriction, born to a 3 gravida mother, out of a second-degree consanguineous marriage, was referred at 82 hours of life with complaints of bilateral scrotal swelling observed on 3rd day of life. On examination, there was a firm and tender swelling over bilateral lower inguinoscrotal region. There was no cry impulse and the testes could not be separately palpated. The neonatologist had recorded the presence of normal testes in the scrotum at birth. Urgent scrotal exploration was performed. The left testis was grayish black and friable with extravaginal torsion (Fig. 1). No improvement was noted after detorsion and warm saline packs. Left orchidectomy was performed. The right testis was also bluish black with torsion (Fig. 1). However, areas of pinkish purple patches appeared after detorsion and warm saline packs; this testis was retained. The immediate postoperative course was uneventful. Biopsy of the left testis revealed immature seminiferous tubules with extensive hemorrhage and necrosis of the lobules.

On 3rd post-operative day, the patient developed a well-demarcated ecchymotic patch over the dorsum of right hand (Fig. 2a) that gradually darkened. Similarly, the right 2nd toe and the left 3rd toe (Fig. 2b) developed bluish purple discoloration. Investigations revealed severe protein C deficiency (<3%) and elevated D-dimer levels. Subsequently, the protein C levels of the father and mother were also found to be low. The mother had a history of cortical vein thrombosis. The ecchymotic patch on the dorsum of the hand and the discoloration of toes gradually disappeared after transfusions of fresh frozen plasma. At follow-up after 2 months, the right testis showed clinical signs of atrophy. Two months later, he developed subcutaneous gangrene of the umbilical region, and the parents were advised to...