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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.

Perinatal 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 3rd 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 late atrophy. Two months later, he developed subcutaneous gangrene of the umbilical region, and the parents were advised to



FIG. 1 Intraoperative image showing the gangrenous left testis (left) and the bluish-black right testis (right).

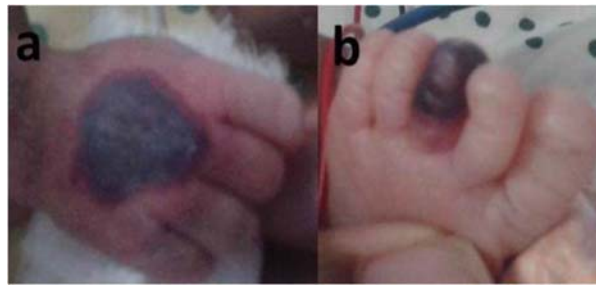


FIG. 2 The skin lesions over the dorsum of the hand (a) and the left third toe (b).

readmit the child. The parents refused, and the child developed spontaneous rupture with bowel evisceration; he died of sepsis at home.

DISCUSSION

Bilateral perinatal testicular torsion in neonates is rare. A large retrospective analysis from one center reported three patients with bilateral torsion in 20 years [2]. Another retrospective case series of 16 neonatal torsions during a 14-year period reported one case of bilateral torsion [4].

Torsion is a well-known cause of an acute scrotum in neonates. Other causes include spontaneous idiopathic hemorrhages, orchitis, epididymitis, and testicular tumor [5]. Large vessel thrombosis involving the renal vein may also present as acute scrotum, mimicking testicular torsion [6].

Purpura fulminans due to protein C deficiency in neonates mainly involves skin and subcutaneous tissues with a predilection for the limbs [3]. It may also present as an acute scrotum due to hematoma [7], and mimic testicular torsion [8]. A hematoma due to protein C deficiency, if diagnosed pre-operatively, may respond to fresh frozen plasma and antithrombotic therapy. Torsion requires an urgent scrotal exploration. However, as seen in our case, the conditions may coexist and present with an acute scrotum with no early skin manifestations.

Timely scrotal exploration is needed because the reported salvage rates are low in postnatal torsion [2]. As detecting testicular blood flow in normal neonates may be difficult [9], it is not advisable to wait for this diagnostic modality. Neonates who undergo scrotal exploration for torsion normally have a smooth post-operative course. However, our patient had a morbid post-operative course due to manifestations of purpura fulminans. Protein C deficiency was diagnosed only after the skin manifestations appeared.

To conclude, bilateral perinatal torsion in neonates is rare and requires urgent scrotal exploration. Severe protein C deficiency may be likely cause, especially if there are skin lesions.

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