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Clinical challenge: a case of testicular tumor disguised as a prenatal testicular torsion

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Abstract

Background: Neonatal tumors account for a minority of prepubertal testicular tumors. Similarly, testicular torsion in the neonate within the intrauterine or postnatal period is a rare event with an estimated incidence of 6 per 100,000. Testicular salvage is almost never achieved in cases of prenatal testicular torsion and there is still no consensus on the ideal management.

Case presentation: A neonate with prenatal testicular torsion that underwent inguinal orchiectomy. On pathological examination, an uncharacterizable, multicystic, and well-delineated testicular tumor was identified. Although rare, testicular tumor presenting as neonatal testicular torsion should be considered in the differential diagnosis of an acute neonatal scrotum.

Conclusions: This case highlights that an inguinal approach for surgical exploration in prenatal testicular torsions may be preferred given the risk, albeit extremely low, of an underlying testicular tumour.

Background

Perinatal testicular torsions are caused by torsion of the spermatic cord occurring before fixation of the tunica vaginalis and dartos within the scrotum. Thus, torsion occurs external to the tunica vaginalis, and is termed "extravaginal". The majority (72% of cases) of perinatal testicular torsion occurs in the prenatal (intrauterine) period but may also occur in the postnatal period (i.e., up to 30 days of life) in 28% of patients [1]. Testicular torsion in the neonate is a rare event, with an estimated incidence of 6 per 100,000 [2]. The incidence is underestimated, as prenatal torsion can cause scrotal nubbins or vanishing testes [2]. Salvage of the testis is almost never achieved in cases of prenatal testicular torsion [2, 3].

The management of perinatal testicular torsion has been the subject of ongoing debate for decades [1]. In a 2008 survey of Canadian pediatric urologists, immediate (29%), or delayed (38–45%) surgical exploration with a scrotal approach and contralateral orchidopexy was favored for prenatal testicular torsion; however, this approach was without consensus [4]. In contrast, a salvage rate of up to 40% has been reported in the context of postnatal torsion; in these cases, all urologists recommended undergoing immediate scrotal surgical exploration [4, 5].

We present the case of a neonate with prenatal testicular torsion that underwent inguinal orchiectomy for extensive necrosis and was subsequently found to have a testicular tumor on pathological examination. This case demonstrates that testicular tumors can present as prenatal testicular torsions. Therefore, an inguinal rather than scrotal approach may be preferred for the surgical exploration of prenatal testicular tumors.

Case presentation

A 36-week gestation baby boy weighing 2920 g was delivered via spontaneous vaginal delivery after a prolonged premature rupture of membranes (PROM) of 26 h. The otherwise healthy 25-year-old mother experienced fever in the peripartum period. The newborn's Apgar score was 9-9 after 1 and 5 min, respectively. Due to prolonged

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PROM and maternal fever, the newborn was placed on amoxicillin and gentamycin. Vital signs remained stable. At 12 h of life, the newborn underwent complete neonatal examination and was found to have a swollen, firm, and bluish left testicle. Sonography of the testis revealed a left swollen testicle measuring 1.9 × 1.7 × 2.3 cm with heterogenous echoes. No color Doppler or arterial pulse Doppler signal was detected. Prenatal torsion of the left testis with necrosis was diagnosed.

The same day, 16 h postpartum, a left inguinal orchiectomy and a right scrotal orchidopexy were performed. The extracted specimen consisted of a 2.5 × 2.0 × 1.8 cm left testicle and was described as dark, large, and necrotic. Upon bivalving the specimen, the surgeon reported widespread hemorrhage and necrosis and evoked the possibility of testicular tumor within the specimen. The newborn had an uneventful post-operative course and was discharged 3 days after surgery in good condition.

Pathological analysis revealed a well-delineated, multicystic, tumor measuring 1.8 cm with necrosis, haemorrhage and calcifications. Extensive haemorrhagic necrosis of the surrounding testicular parenchyma with dystrophic calcifications, consistent with subacute/remote testicular torsion was reported (Fig. 1). Immunohistochemistry could not be performed on the specimen due to the absence of residual viable tissue. The differential diagnosis of the mass reported by the pathologist comprised of germ cell tumors (e.g., teratoma, yolk sac tumors), juvenile granulosa cell tumor (JGCT) and hemangioma.

At one month of life, chest radiography was negative for metastasis and serum tumor markers (STM) were within physiological range for a newborn (AFP 2030 [up to 80,000]; BHG < 2 [0–4.9]; LDH 635 [470–920]). The case was presented at our institution's tumor board, which recommended initial monitoring every 3 months with serum AFP and b-HCG and sonography. At 12 months, no evidence of recurrence was found and follow-up was performed every 6 months for 2 years and then annually. Adjuvant chemotherapy was not deemed necessary. After 15 years of follow-up, no recurrence was detected and STM remained normal.

Discussion

Prepubertal testicular tumors represent 1% of solid pediatric tumors [6]. The epidemiology varies from that of adult testicular tumors: yolk sac tumors and teratoma, both nonseminomatous germ cell tumors, comprise 85% of testicular tumours in boys younger than 12 years, while representing 1% in adults [7]. In neonates younger than 30 days, 22 cases, or 6.5% of all prepubertal testicular tumors, have been reported in the *Prepupertal Testis Tumour Registry* with the following distribution: 6 (27%) yolk sac tumors, 6 gonadal stromal tumors, 6 JGCTs (sex-cord stromal tumors), 2 (9%) gonadoblastomas (mixed germ cell and stromal tumors), 1 (5%) teratoma, and 1 hamartoma [8]. Of these tumors, 7 (31%) presented at birth as a testicular mass.

In this case, knowing an orchiectomy was the most likely outcome based on sonographic findings, the surgeon opted empirically for an inguinal approach. To our knowledge, only one other similar case of a testicular tumor (teratoma) presenting as a prenatal testicular torsion in a baby with descended testes has been reported [9]. An inguinal approach is optimal if the surgeon proceeds to surgery for a prenatal testicular torsion, given the chance of an underlying testicular tumor.

Unfortunately, we could not achieve a histopathological diagnosis as extensive testicular necrosis rendered analysis impossible. However, the favorable evolution of the patient documented over 15 years is highly suggestive of a benign testicular mass.

Heterogenous echoes, occasional calcifications, and multiple intratesticular cysts are frequent sonography findings in teratoma but have also been associated with testicular cysts, JGCT, and cystic dysplasia of the rete testis [10]. Moreover, after torsion, cystic degeneration with formation of cystic spaces filled by necrotic debris and haemorrhagic infarction can produce similar images [10]. Mature teratomas have a median age of presentation of 13 months, are benign tumors and do not express AFP [10]. No teratoma, in the absence of immature components, has been documented to cause metastasis [11]. Thus, mature teratoma may be treated with partial orchiectomy [12]. Elevated AFP is suggestive of yolk sac tumors, which are the most common malignant prepubertal testicular tumors, generally presenting at 16 months [10]. Yolk sac tumors require inguinal orchiectomy, close follow-up, and may be considered for platinum-based chemotherapy according to the stage [13]. Seven-year survival of children younger than 2 years old presenting with stage I yolk sac tumor is 90% [13]. In children younger than 1 year, AFP is of lesser diagnostic value in distinguishing mature teratomas from yolk sac tumor since there is physiological AFP production in the liver and gastro-intestinal tract, as well as by the fetal yolk sac [7]. Finally, JGCTs are benign sex-cord stromal tumors. Most JGCTs (90%) are diagnosed in infants < 6 months and produce cystic lesions [10, 14]. The only stromal tumors that have been documented to cause metastasis in neonates are undifferentiated stromal tumors. These rare tumors require close follow-up, radical inguinal orchiectomy, and may benefit from retroperitoneal lymph node dissection, radiation, or chemotherapy [15].

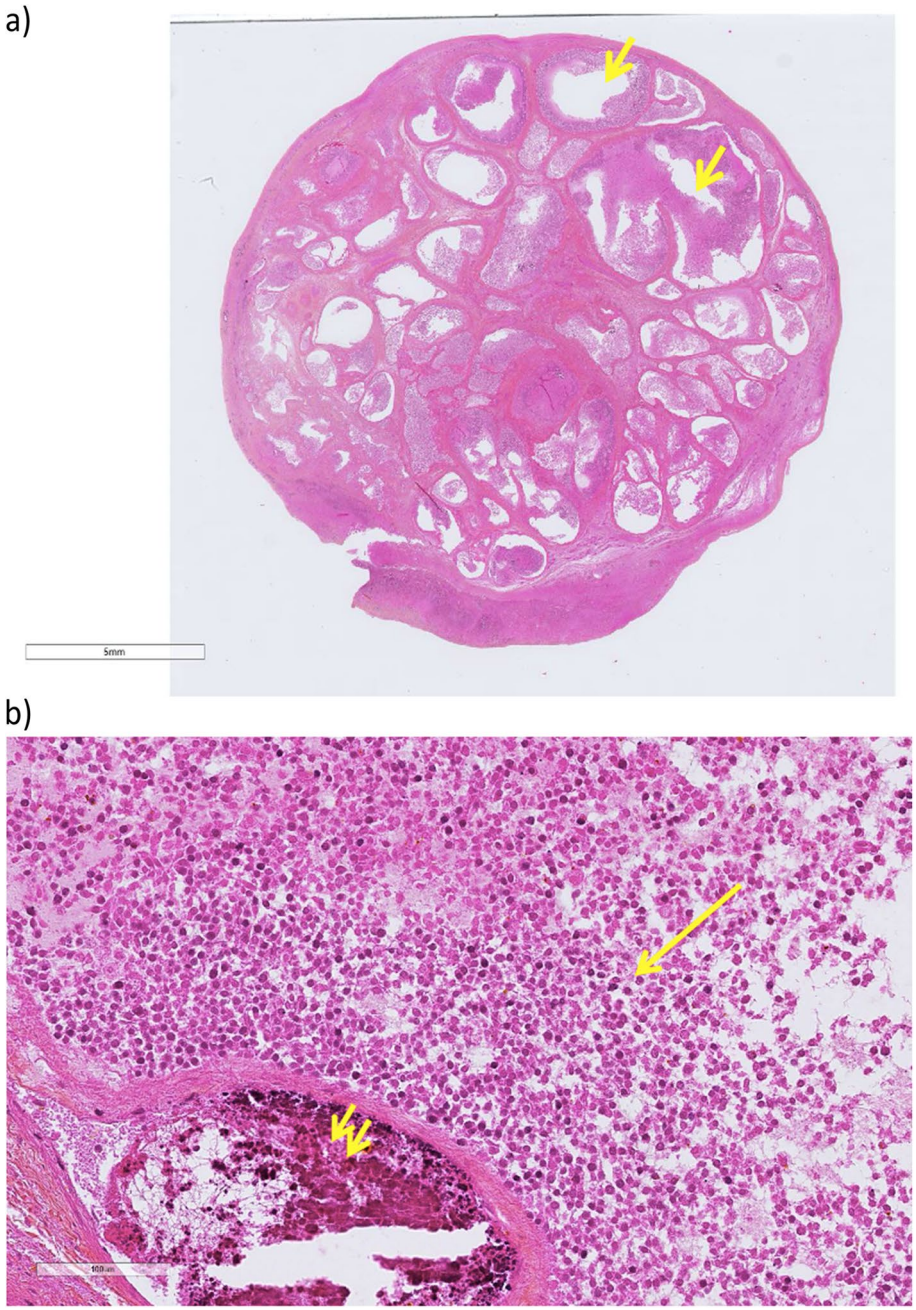

Fig. 1 **a** Necrotic tumor with cystic degeneration (single arrows). **b** Prominent necrotic small round blue cell tumor (single arrow) with dystrophic calcification (double arrow)

Conclusion

Although rare, testicular tumors presenting as prenatal and postnatal testicular torsions should be considered in the differential diagnosis of an acute neonatal scrotum. Here, we present a case of a testicular tumor that presented as prenatal testicular torsion. The mass was not detected on sonography and the neonate underwent immediate inguinal orchiectomy with a pre-operative diagnosis of necrotic testicular torsion, without any suspicion of tumor. If surgical exploration is undertaken for a prenatal testicular torsion, the surgeon may consider an inguinal rather than scrotal approach, given the possibility of an underlying testicular tumor.

Abbreviations
JGCT: Juvenile granulosa cell tumor; PROM: Premature rupture of membranes; STM: Serum tumor markers.

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Competing interests
The authors declare that they have no competing interests.

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References

1. Das S, Singer A. Controversies of perinatal torsion of the spermatic cord: a review, survey and recommendations. J Urol. 1990;143:231–3.
2. John CM, Kooner G, Mathew DE, Ahmed S, Kenny SE. Neonatal testicular torsion-a lost cause? Acta Paediatr. 2008;97:502–4.
3. Brandt MT, Sheldon CA, Wacksman J, Matthews P. Prenatal testicular torsion: principles of management. J Urol. 1992;147:670–2.
4. Guerra LA, Wiesenthal J, Pike J, Leonard MP. Management of neonatal testicular torsion: Which way to turn? Can Urol Assoc J. 2008;2:376–9.
5. Sorensen MD, Galansky SH, Striegl AM, Mevorach R, Koyle MA. Perinatal extravaginal torsion of the testis in the first month of life is a salvageable event. Urology. 2003;62:132–4.
6. Kay R. Prepubertal testicular tumor registry. Urol Clin North Am. 1993;20:1–5.
7. Ross JH, Rybicki L, Kay R. Clinical behavior and a contemporary management algorithm for prepubertal testis tumors: a summary of the Prepubertal Testis Tumor Registry. J Urol. 2002;168:1675–9.
8. Levy DA, Kay R, Elder JS. Neonatal testis tumors: a review of the Prepubertal Testis Tumor Registry. J Urol. 1994;151:715–7.
9. Führer S, May M, Koch A, Marusch F, Gunia S, Erler T, et al. Perinatal/neonatal torsion of a testicular teratoma: a case report. J Perinatal. 2005;25:220–2.
10. Garrett JE, Cartwright PC, Snow BW, Coffin CM. Cystic testicular lesions in the pediatric population. J Urol. 2000;163:928–36.
11. Metcalfe PD, Farivar-Mohseni H, Farhat W, McLorie G, Khoury A, Bägli DJ. Pediatric testicular tumors: contemporary incidence and efficacy of testicular preserving surgery. J Urol. 2003;170:2412–6.
12. Sugita Y, Clarnette TD, Cooke-Yarborough C, Chow CW, Waters K, Hutson JM. Testicular and paratesticular tumours in children: 30 years' experience. Aust N Z J Surg. 1999;69:505–8.
13. Liu HC, Liang DC, Chen SH, Liu FL, Chang PY, Sheu JC, et al. The stage I yolk sac tumor of testis in children younger than 2 years, chemotherapy or not? Pediatr Hematol Oncol. 1998;15:223–8.
14. Kao CS, Cornejo KM, Ulbright TM, Young RH. Juvenile granulosa cell tumors of the testis: a clinicopathologic study of 70 cases with emphasis on its wide morphologic spectrum. Am J Surg Pathol. 2015;39:1159–69.
15. Thomas JC, Ross JH, Kay R. Stromal testis tumors in children: a report from the prepubertal testis tumor registry. J Urol. 2001;166:2338–40.

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