Neonatal testicular tumour presenting as an acute scrotum

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Juvenile granulosa cell tumour (JGCT) is a rare benign stromal cell tumour of the testis accounting for approximately 1% of all paediatric testicular tumours. Presenting primarily as a painless testicular mass, the tumour may be associated with undescended testis, hydrocele or testicular torsion. Abnormal karyotype has also been described. We describe an unusual case of a neonatal juvenile granulosa cell tumour presenting as an acutely swollen, tender testis, originally diagnosed as an acute hydrocele. We describe the management and review the literature pertaining to this rare differential diagnosis of a neonatal acute scrotum. *Ann Pediatr Surg* 8:19–21 © 2012 Annals of Pediatric Surgery.

Annals of Pediatric Surgery 2012, **8**:19–21

Keywords: acute scrotum, granulosa cell tumour, neonatal, testicular tumour

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Received 2 August 2011 **accepted** 22 November 2011

Case report

A term infant showed signs of discomfort due to a left hemiscrotal swelling and inguinal erythema at 3 weeks of age. His local hospital carried out a diagnosis of hydrocele. At routine postnatal checkup the testis was described as lying at the pubic tubercle, but was otherwise normal.

He failed to settle and was referred at 4 weeks of age as a case of acute scrotum or hernia. An examination demonstrated left scrotal enlargement with a lax hydrocele surrounding an enlarged, irregular left testis with marked oedema and inflammation over the inguinal canal.

The ultrasound scan (USS) demonstrated a vascular heterogeneous hyperechoic left testis (left testis: $15 \times 15 \times 24$ mm, right testis: $5 \times 5 \times 12$ mm) with several well-defined cystic pockets (Figs 1–3). Full blood count, electrolytes and urine analyses were normal. Serum beta-human chorionic gonadotrophin (β-HCG) and alpha fetoprotein (α-FP) were within normal limits for age, at less than 3 mIU/ml and 1387 ng/ml, respectively.

Although the history of discomfort with inguinal oedema might have suggested a possible resolving neonatal torsion or an inflammatory condition, testicular tumour was considered likely because of the result on the USS, and a high approach exploration was performed the following day. Findings on surgery were of a homogeneously enlarged testis with an oedematous cord, with no torsion or evidence of testicular ischaemia. Altered blood was noted to be present within the tunica, patent processus vaginalis and peritoneum, although there was no evidence of intra-abdominal bleeding.

Macroscopically, the testis showed almost entirely a cystic and solid appearance with yellow patches and small areas of haemorrhage, with only a small peripheral layer of normal-appearing testicular tissue. The capsule surrounding the testis was intact.

Microscopically, the testis appeared to be composed of a lobular structure with follicular-like spaces, with focal cystic degeneration and some spaces containing blood (Figs 4 and 5). The cells were negative for α-FP, β-HCG and placental alkaline phosphatase. Immunohistochemistry confirmed positivity of the cells with inhibin and cytokeratin. This was consistent with a diagnosis of a juvenile granulosa cell tumour with bleeding into the patent processus vaginalis causing an acute presentation with discomfort and inguinal inflammation.

Follow-up in the outpatient clinic until 24 months postoperatively showed no recurrence and an appropriate fall in α-FP.

Discussion

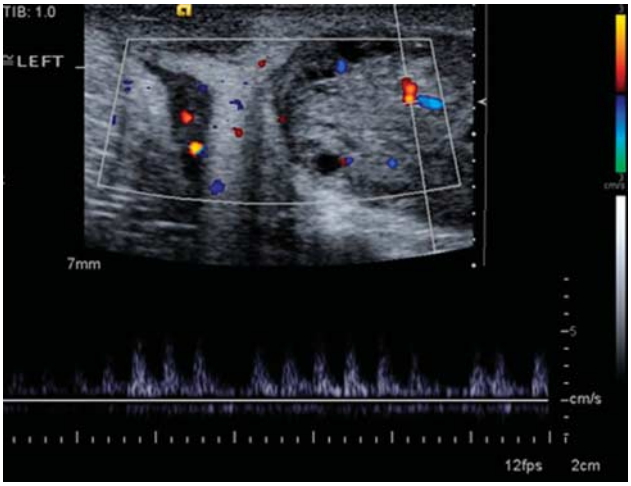
Testicular tumours are rare and account for approximately 2% of all paediatric solid tumours, occurring with an estimated frequency of 0.5 : 100 000 children [1–3]. One subset of these is the stromal tumours representing 8% of all testicular tumours, which can be subdivided into granulosa cell tumour Juvenile granulosa cell tumour (JGCT), Leydig cell tumour and Sertoli cell tumour [3]. Although isolated cases have been identified in adults [4] and on antenatal USS [5], the majority occur within the first year of life. Although JGCT constitutes only a tiny percentage of testicular tumours, it is the most common testicular tumour in children less than 6 months of age, accounting for 27% of all cases [3,6]. A literature review of 36 reported cases of JGCT [7] found that all cases but one were identified in children below the age of 1 year.

JGCT is associated with ambiguous genitalia both in XO/XY mosaics and XXY triploidy [8,9] and has been associated with neonatal undescended testes [7] and neonatal torsion [10]. There is a related hydrocele in approximately 10% of cases [11,12] Endocrine features are not usual in the presentation of JGCT and can be used as a means of exclusion for this diagnosis [10].

Definitive preoperative diagnosis remains difficult; assessment includes normal α-FP and β-HCG and characteristic USS, which usually demonstrates multicystic, discrete lesions within the parenchyma of the testis. [5]. Preoperative diagnosis suggests the possibility of testis-sparing surgery for some cases [3,10] because of the benign nature of JGCT of the testis, with no patients

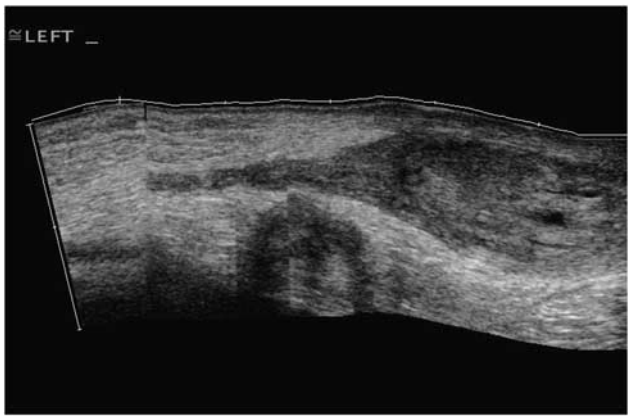
1687-4137 © 2012 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000409920.48006.a5

Fig. 1



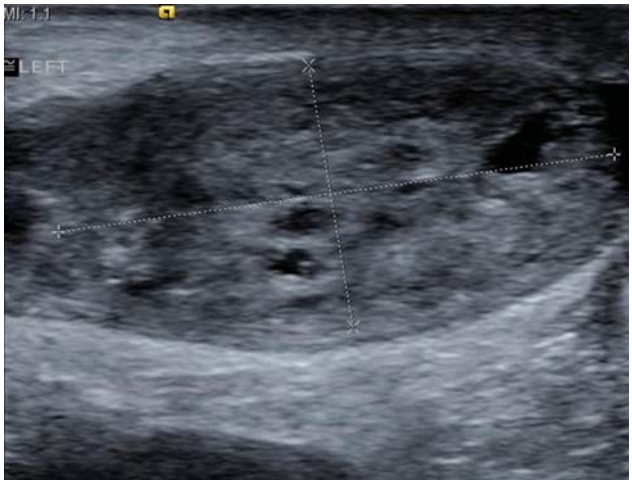
Ultrasonogram: showing good arterial and venous flow within the substance of the testis – the degree of blood flow given the degree of parenchymal abnormality made previous torsion a less likely diagnosis.

Fig. 2



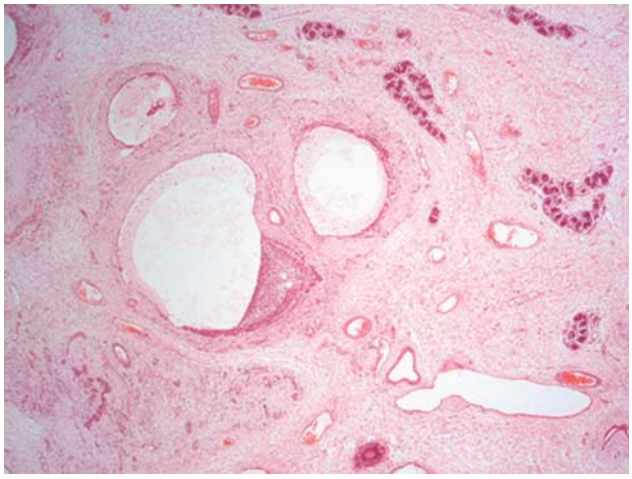
Ultrasonogram: vascular heterogeneous hyperechoic left testis, with several well-defined cystic pockets.

Fig. 3



Ultrasonogram: enlarged left testis (15 × 15 × 24 mm) – heterogeneous and hypoechoic – with several well-defined cystic areas within the testis. A small associated hydrocele.

Fig. 4



Histopathogy: 1 × 200 magnification: showing a cystic space lined by granulosa cells with remnants of testicular tubules adjacent.

reported with metastatic disease at the time of presentation or at follow-up. Most centres still perform full orchidectomy, but partial orchidectomy or enucleation, particularly in neonates of less than 3 months fulfilling the above criteria, has been advocated when there is preserved normal testis [10]. This would not have been a viable option in this case as there was very little normal testicular tissue present.

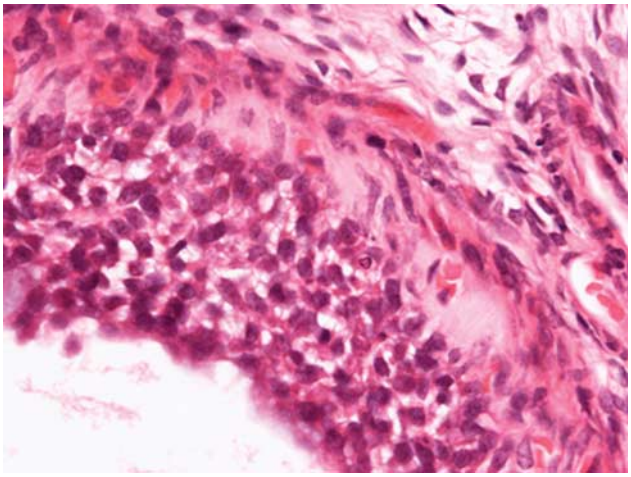
Definitive diagnosis relies on cellular pathology with identification of morphological features that are difficult to distinguish from those of yolk sac tumours; each has multicystic areas containing mucin intermixed with solid components. When microscopic appearances are equivocal, a positive response to inhibin immunohistochemical staining, although remaining negative to staining for α-FP, is a sensitive and now widely used method for accurately diagnosing JGCT [13]. Recent studies have suggested that trisomy 12 may be widespread (75%) in paediatric granulosa stromal cell tumours and can be identified by fluorescence in-situ hybridization on paraffin-embedded material [14]

Although metastasis has not been described in neonates, close postoperative follow-up is recommended particularly in those cases with tumours that have high mitotic figures and are less well differentiated. [3].

Conclusion

In neonates who present with an enlarged scrotum, JGCT is an important, albeit rare, differential diagnosis. The majority present as mass or torsion, and USS and tumour

Fig. 5



Histopathology: 2 × 400 magnification: showing cells lining the cystic spaces with granulosa cell morphology.

markers can aid preoperative diagnosis. The case presented here unusually mimicked an acute scrotum probably because of the intravaginal haemorrhage identified during operation. An appropriate course of investigation and operative approach was possible because of efficient USS and awareness of the potential diagnosis of JGCT. Radical or partial orchidectomy is the current recommended treatment with no further systemic management necessary because of its benign course. Careful follow-up, however, is still recommended because of the rarity of this condition and because of our limited knowledge of its clinical course.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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