A rare testicular solid mass in children: juvenile granulosa cell tumour of testis

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Abstract

Juvenile granulosa cell tumour (JGCT) of the testis is a benign neoplasm rarely seen in children. It usually presents as a unilateral scrotal mass and can be associated with genital ambiguity and chromosomal anomalies. Radical orchiectomy is the treatment of choice. We present an infant with a JGCT of the testis and we review the typical findings of the disease.

Introduction

Prepubertal testicular tumours are rare, accounting for 1% of all pediatric solid tumours. Sex cord-stromal tumours represent 8% of those prepubertal testicular tumours.[1] Granulosa cell tumours of the testis are a subtype of sex cord-stromal tumours. These rare neoplasms are morphologically similar to their ovarian counterparts and can be distinguished by adult or juvenile type. Even though juvenile granulosa cell tumour (JGCT) of the testis only accounts for 1% to 5% of all prepubertal testis tumours,[2] it is one of the most common neoplasms of the testis in the first 6 months of life.[3] The first case of JGCT of the testis was reported by Crump in 1983.[4] Less than 50 cases have been reported,[5] all with a benign course.

After obtaining approval by the research ethical committee at our institution, we present a 3-month-old male with JGCT and typical features of the disease.

Case report

A 3-month-old male was referred to our pediatric urology clinic with a right testis that was enlarged for a month. His physical examination was unremarkable except for a hard mass to the right testis. It was mobile and painless. There was no fever, no digestive symptoms, no sign of infection and no lymphadenopathy. The penis and left testis were normal. The child was born with a normal birth weight, at term, without prenatal or postpartum medical problems.

Testicular ultrasonography revealed a near-complete replacement of the right testis by a 3.7 × 2.2 × 2.5 cm multicystic mass without any vascularization demonstrated by Doppler sonography (Fig. 1a). The left testis was normal. His serum alphafoetoprotein and β-HCG values were within normal limits for his age. His karyotype was 46 XY.

He underwent right radical inguinal orchiectomy. Marcoscopic evaluation revealed a 4.1 × 2.8 × 2.2 cm testis with a unilocular cystic mass containing pinkish fluid. Microscopic examination revealed a unilocular cystic mass bordered by many sheets of cells with oval nuclei and abundant cytoplasm (Fig. 1b). Mitoses were rare. Immunohistochemically, cells were positive for inhibin (Fig. 2a), vimentin, smooth muscle actin, protein S-100 and CD99 and negative for placental-like-alkaline-phosphatase (PLAP) and CD117, which are two markers of germ cell tumours (Fig. 2b). The pathology report confirmed the diagnosis of JGCT of the testis.

Discussion

JGCT of the testis is a rare, benign, subset of sex cord-stromal tumours which also include Leydig cell, Sertoli cell and undifferentiated cell tumours.[6] In the ovary, the adult form is much more common than the juvenile form; in the testis, the juvenile form is more common than the adult one.[7] The JGCT of the testis has a tendency to occur in very young patients, mostly in the first 6 months of life.[7] Ninety percent are found in infants of less than a year old.[1]

It typically presents as a painless unilateral scrotal mass. The differential diagnosis for solid testicular mass at this age also includes teratomas, epidermoid cysts, yolk sac tumours (endodermal sinus tumours) and mixed germ cell tumours.[11] Only two cases of bilateral JGCT of the testis have been reported, both in intraabdominal testis.[8,9] In 20% of the

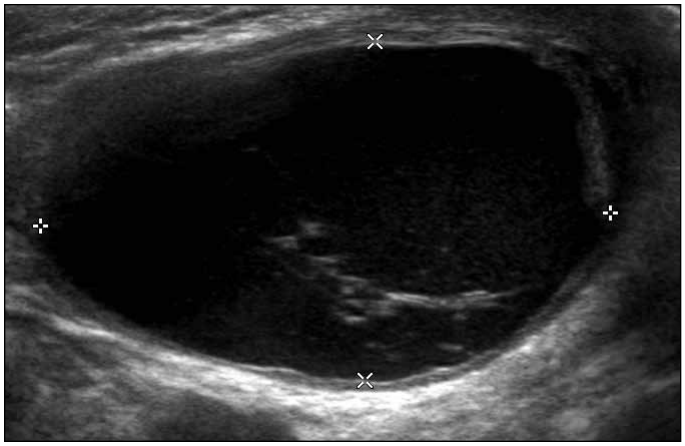

Fig. 1a. Ultrasonography revealing a near-complete replacement of the right testis by the tumour.

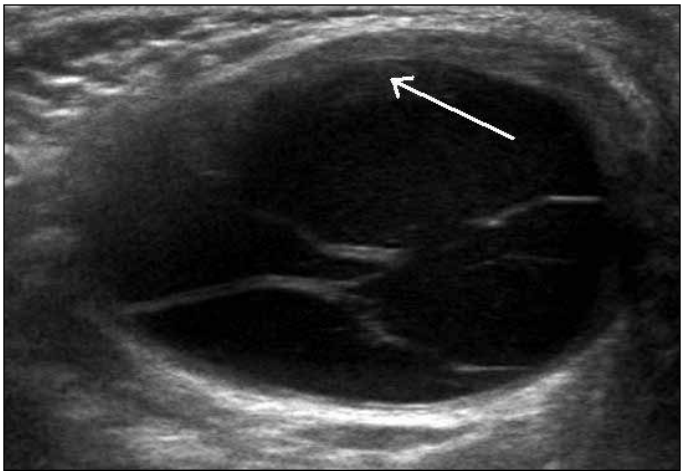

Fig. 1b. Ultrasonography of the right testis showing a cystic mass with septations. A rim of testicular tissue was present (arrow).

reported cases of JGCT of the testis, chromosomal anomalies have been detected, such as structural abnormalities of the Y chromosome and XO/XY mosaicism,[10] and some had genital ambiguity. The karyotype is only indicated after obtaining the final pathology report because this particular tumour has been reported to be associated with chromosomal anomalies. No endocrine manifestations, such as isosexual pseudoprecocity or estrogenic manifestations, were present.[1] The tumour markers are usually normal.[1] Our patient had normal serum alphafoetoprotein (AFP) and β-HCG values. Since it is known that the most common malignant tumour in children is yolk sac tumour and that this tumour does not elaborate β-HCG, AFP is the only important tumour marker in prepubertal patients. Indeed, 90% of yolk sac tumours in children have elevated AFP. However, an important caveat in the use of AFP is that serum AFP is normally high in infancy and does not decrease to normal adult levels until nearly the child is 1 year old. Therefore, while an elevated AFP in a child over 1 year old with a testis tumour almost always reflects the presence of a yolk sac tumour, an elevated level in infants can occur in the setting of a benign tumour.[11] It is also useful to mention that yolk sac tumour of the testis in young boys peaks after 6 months of age, which is after the peak of JGCT.[7]

Because JGCT of the testis is rare, a description of imaging findings is limited. Testicular ultrasonography usually reveals a well-defined, large multicystic intratesticular solid mass.[10] Ultrasonography features can be helpful to distinguish a benign tumour from a malignant one. Benign tumours tend to be well-circumscribed with sharp borders and decreased blood flow on Doppler studies, as in the case with our patient.[11] Metastatic evaluation can be deferred until after the tumor is excised for patients who likely harbour a benign tumour.[11] In our case, it was not necessary to perform an extensive workup before the pathological diagnosis. It is well-known that about 75% of testis tumours in prepubertal patients are benign, a computed tomography scan should only be performed in children once a malignant diagnosis has been documented.[11] The most common sites for metastases of testis tumours are lungs and the retroperitoneum.[11]

On gross pathology, JGCT of the testis typically appears as a tan to yellowish mass with a mixture of solid and cystic regions without necrosis or hemorrhage.[1] Microscopically, cysts are lined with single or multiple layers of granulosa cells with round to oval nuclei that are hyperchromatic with eosinophilic cytoplasm. Mitoses may be prominent and numerous.[1] Granulosa cells stain positive for vimentin, inhibin, pancytokeratine, smooth muscle actin and protein S-100.[12]

JGCT of the testis are considered borderline tumours; no locally advanced, metastatic or relapsed cases have been reported.[6] Management includes radical inguinal orchiectomy with close surveillance, including chest radiographs and abdominopelvic ultrasonography of the retroperitoneum.[1] It is important to note that the benign outcome of JGCT of the testis suggests a role for testis-sparing surgery when healthy testis tissue is present. Indeed, tumor enucleation with no metastatic evaluation could be sufficient.[10-11] However, because of the confusion about whether this tumour is benign or borderline, the usual practice, which is the one adopted in our centre, remains radical inguinal orchiectomy.

Conclusion

JGCT of the testis should be kept in mind in the differential diagnosis of newborns and children scrotal masses, especially in the first year of life. Reporting the cases and giving facts that will help to make an adequate differential diagnosis are needed to improve diagnosis and therapy options.

Competing interests: None declared.

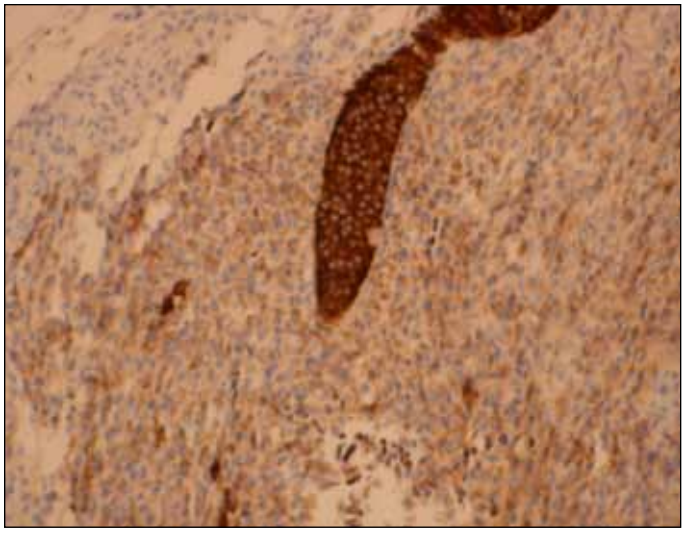

Fig. 2a. Juvenile granulosa cells are typically positive for inhibin.

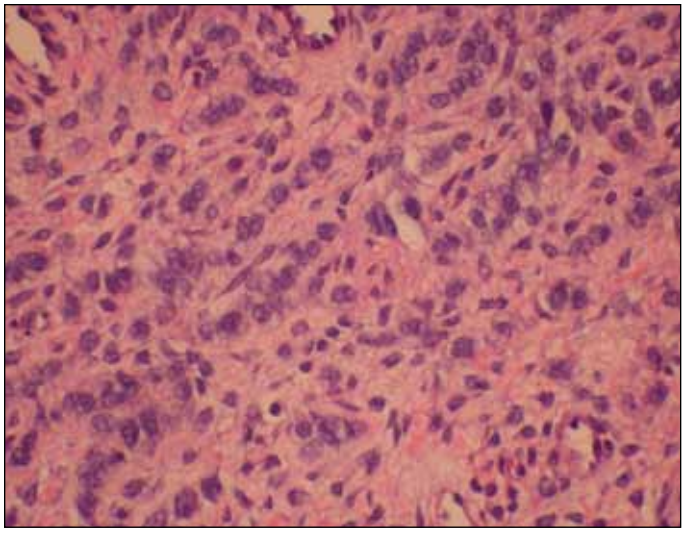

Fig. 2b. Microscopic appearance of a juvenile granulosa cell tumour: many layers of granulosa cells with oval nuclei and abundant cytoplasm.

This paper has been peer-reviewed.

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