

Congenital juvenile granulosa cell tumor of the testis: Case report and literature review

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Abstract

Juvenile granulosa cell tumor (JGCT) is a very rarely diagnosed benign tumor, accounting for 1.2% of all prepubertal testicular tumors. A full-term healthy neonate was diagnosed with a painless left scrotal mass. During evaluation it was identified to have about two times the volume of the contralateral testis, presenting a firm consistency, not as hard as the consistency of a prenatal testicular torsion. Doppler ultrasound detected a multicystic left testicular mass, with normal blood flow, but failed in detecting normal-appearing testis. Human chorionic gonadotropin (β -HCG) and serum alpha-fetoprotein (AFP) were normal. Inguinal approach was performed, section of the lesion was sent to frozen biopsy and excluded yolk sac tumor, and however the impossibility of detecting normal testis tissue indicated orchiectomy with high ligation of the spermatic cord. Histological evaluation demonstrated gray testicular parenchyma with multicystic aspect fulfilled with yellow fluid. The usual clinical presentation of JGCT is a painless scrotal mass, radiological imaging demonstrates a multicystic tumor. Tumoral markers levels are normal and the standard treatment is the inguinal orchiectomy.

Keywords

Granulosa cell tumor; testis tumor; scrotal mass.

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Accepted for publication: 22 April 2016

Introduction

Prepubertal testicular tumors are rare and account for 1% of all pediatric solid tumor [1]. Juvenile granulosa cell tumor (JGCT) is a benign neoplasm that represents the most common stromal cord tumor of the infant testis, accounting for 1.2% of all prepubertal testicular tumors [2]. The usual clinical presentation is a painless scrotal mass, detected during clinical routine examination or perceived by the parents. Radiological imaging demonstrates a multicystic circumscribed tumor [2,3]. Tumoral markers levels are normal and the standard treatment is the inguinal orchiectomy [4]. It has only been clearly delineated as a distinct entity since 1985 and has been named so due to its histological similarity to the ovarian granulosa cell tumor [5]. We report a congenital case of this rare tumor, diagnosed in the first day of life.

Case Reports

A full-term health neonate was diagnosed with a painless left scrotal mass. During evaluation it was identified to have about two times the volume of the contralateral testis, presenting a firm consistency, not as hard as the consistency of a prenatal testicular torsion [Fig. 1].

Doppler ultrasound detected a multicystic left testicular mass, with normal blood flow,

but failed in detecting normal-appearing testis [Fig. 2]. The left testis was also bigger than the right measuring 2.66x1.03x1.81 cm with volume 2,60ml.



Fig. 1. Clinical evaluation.



Fig. 2. Ultrasound: multicystic left testis.

Human chorionic gonadotropin (β -HCG) and serum alpha-fetoprotein (AFP) were normal. Inguinal approach was performed, exposing the testis and spermatic cord. After cord clamping, a section of the lesion was sent to frozen biopsy, excluding yolk sac

tumor. However the impossibility of detecting normal testis tissue indicated orchiectomy with high ligation of the spermatic cord [Fig. 3].



Fig. 3. Surgical aspect.

Histological evaluation demonstrated gray testicular parenchyma with multicystic aspect fulfilled with yellow fluid [Fig. 4]. Postoperative evolution was uneventful. Two years after surgery the patient is asymptomatic and being followed by pediatric oncology.

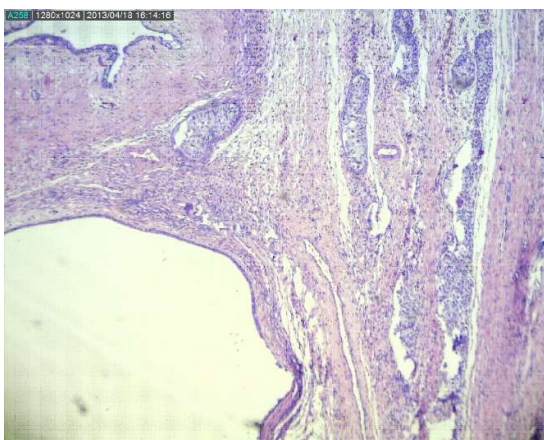


Fig. 4. Histological evaluation.

Discussion

Stromal testicular tumors account for 8% of testicular neoplasms [5]. JGCT of the testis is a rare, benign, subset of sex cord-stromal tumors which also include Leydig cell, Sertoli cell and undifferentiated cell tumor. They represent 10-20% of all sex cord-stromal tumors¹. Ninety percent are found in infants of less than a year old, mostly in the first six months of life [6]. They present as painless scrotal or abdominal masses, more often affecting the left testis. The differential diagnosis for solid testicular mass at this age also includes teratoma, epidermoid cysts, yolk sac tumors and mixed germ cell tumors [2].

There are no demonstrable hormonal effects in most cases [4]. Tumor markers such as alpha-fetoprotein and β -HCG are within the normal for this age. A total of 20% of the reported cases are children with chromosomal abnormalities affecting the Y chromosome [5] and can also be associated with mosaicism and ambiguous genitalia [7]. Testicular ultrasonography usually reveals a well-defined large multicystic intratesticular solid mass [2]. On gross pathology it appears as a tan to yellowish mass with a mixture of solid and cystic regions that are lined with single or multiple layers of granulosa cells with round to oval nuclei that are hyperchromatic with eosinophilic

cytoplasm [8]. The granulosa cells stain positive for cytokeratin and vimentin [5].

Only a small percentage of JGCT originate from undescended testis, however the literature shows that there are no difference in behavior [7].

JGCT of the testis are considered borderline tumors, no locally advanced, metastatic or relapsed cases have been reported [9]. The inguinal orchiectomy still remains the cornerstone in the treatment of this entity [5]. Due to the benign outcome a testicular sparing surgery has been recently proposed, but it is still not general practice [3,9]. There is compelling evidence to suggest that this option is viable specially in cases without any evidence of metastases at presentation, normal levels of serum tumor markers and

small juvenile tumors [4,9]. They usually present excellent prognosis [10].

Conclusion

Juvenile granulosa cell tumors are a rare entity that has an important role as a differential diagnose of testicular masses. The case reported at this article shows a classic presentation of JGCT. As they are uncommon reporting the cases is needed for discussing diagnosis and therapy options

Acknowledgements

The author(s) declare that they have no competing interests and financial support

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