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Juvenile Granulosa Cell Tumor of the Testis in a Newborn with Swelling in the Scrotum: A Rare Case and Literature Review

CASE REPORT

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ABSTRACT

Juvenile granulosa cell tumors (JGCTs) of the testis in newborns are very rarely seen benign tumors. They occur in association with sex chromosome disorders and ambiguous genitalia. Alpha-fetoprotein levels have a poor diagnostic value for teratomas in infants <6 months of age. Most of JGCTs are confused with a teratoma based on histopathological analysis results. Inguinal orchiectomy yields satisfactory treatment results. We present the case of our patient diagnosed with a JGCT of the testis and review the relevant literature.

Keywords: Juvenile granulosa cell tumor, testis, newborn, orchiectomy

INTRODUCTION

Testicular tumors comprise 1% of all pediatric tumors and are rarely seen in newborns. Granulosa cell tumors, which are a sex cord-stromal tumor group, can be divided into adult granulosa cell tumor and juvenile granulosa cell tumor (JGCT) subtypes (1). In contrast to adult granulosa cell tumors, the juvenile subtype has benign characteristics. A physical examination usually reveals a painless mass, and inguinal orchiectomy is sufficient for treatment (2). This study aimed to present the rare case of a patient diagnosed as having JGCT and to review the relevant literature.

CASE REPORT

A 12-day-old male infant was brought to our clinic with swelling in the left scrotum that was noticed by the family 1 week prior. A physical examination revealed a palpable, but painless, mobile, and diffuse, 2×2 cm mass with a smooth surface. The right testis had dimensions of 7×6 mm, and penile and other system examinations were unremarkable. There were no changes in the color of the scrotum (Figure 1). A complicated cystic lesion, the contours of which could not be discriminated from the left testicular parenchyma, contained 15×10 mm septations, and occasional areas of vascular flow were detected on performing scrotal ultrasound (US; Philips, Epiq 5, USA) (Figure 2a, b). A 2 mm simple cyst was observed in the right epididymis. Pelvic magnetic resonance imaging (MRI; Philips, Ingenia 1,5T, USA) revealed a septated mass lesion in the left testis with regular contours, which could not be differentiated from the left testicular parenchyma, containing a 15×11 mm cystic area and millimeter-sized cysts in the head of the right epididymis (Figures 3a, b). The levels of human chorionic gonadotropin (hCG; <2 mIU/mL) and alpha-fetoprotein (AFP; 33.118 ng/mL on day 7 and 2.209 ng/mL on day 25) were also measured. Chromosomal analysis revealed a 46 XY genotype. The infant underwent high scrotal orchiectomy through a left inguinal incision. The orchiectomized testis measured 2×1.5 cm. Three cysts with regular contours were de-

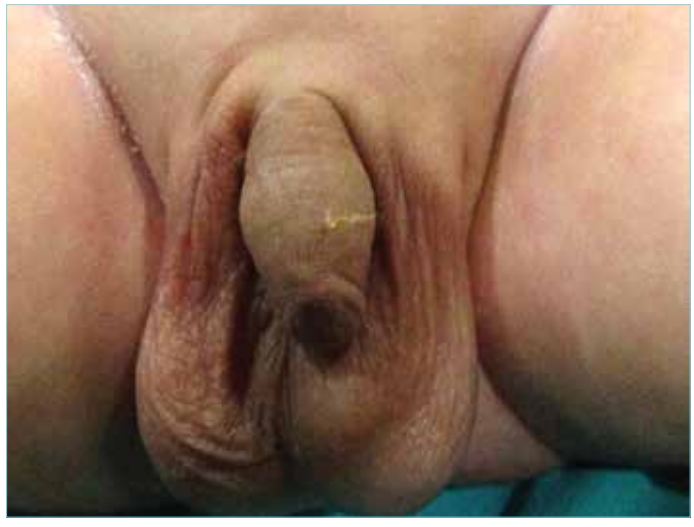

Figure 1. **Image of the scrotum at initial examination**

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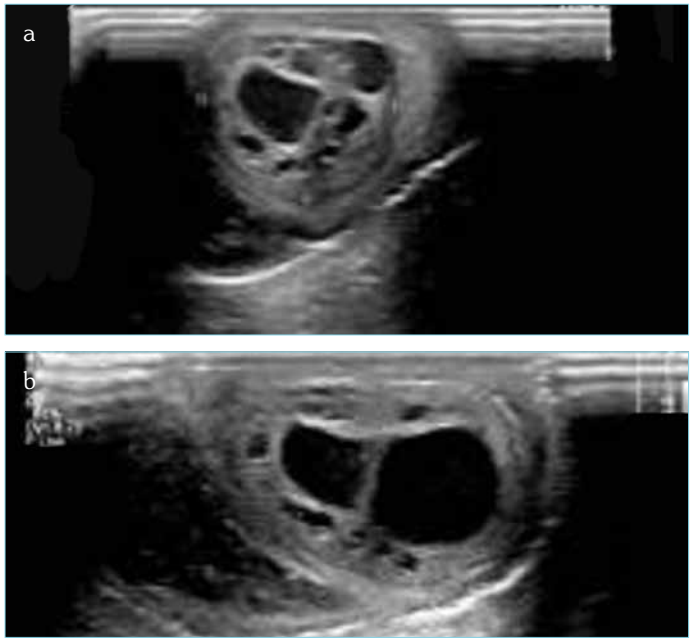

Figure 2. a, b. Ultrasound image of the left testis scrotum. A complicated cystic lesion (teratoma), whose contours could not be differentiated from the left testicular parenchyma, contained 15×10 mm septations

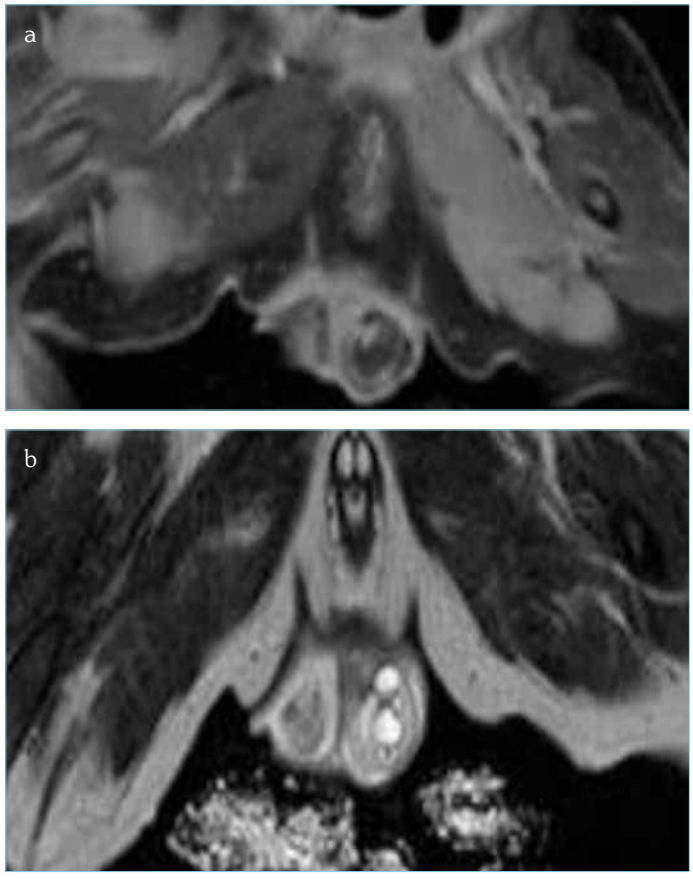

Figure 3. a, b. Contrast, T1A coronal section(a) and T2A coronal section (b); Magnetic resonance image of the left testis scrotum. Mass lesion with a regular contour, cystic area, and areas of septal contrast uptake measuring 15×11 mm

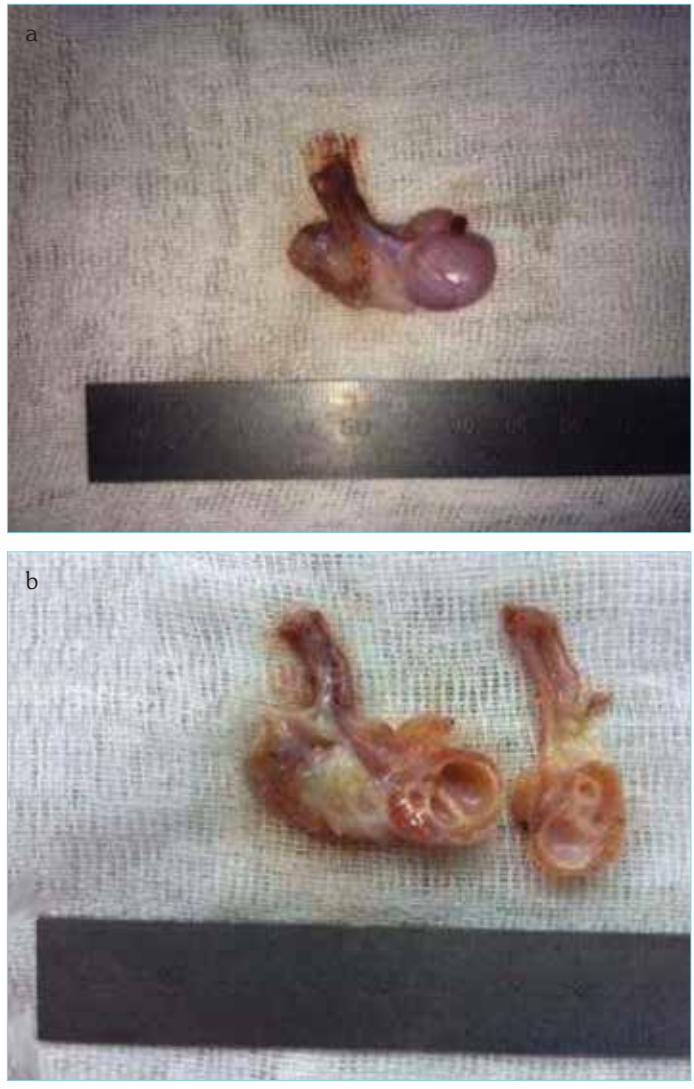

Figure 4. a, b. Macroscopic appearance of the orchiectomy material

tected in sections, and these were surrounded with a dirty white fibrous capsule containing yellow serous fluid; the largest cyst was 1 cm in diameter (Figure 4a, b). A histopathological examination of the mass revealed a JGCT. The postoperative AFP level was 450 ng/mL. No complications were encountered during the follow-up period.

DISCUSSION

Granulosa cell tumors belong to the sex cord-stromal tumor group and are divided into adult granulosa cell tumor and JGCT subtypes (1). Adult granulosa cell tumors are malignant and generally originate from the ovaries, whereas JGCTs are benign and originate from testicular tissue (3).

Although JGCTs are rare testicular tumors, they are the most frequently seen testicular tumor in infants <6 months of age (1). The most commonly seen clinical finding is unilateral, painless scrotal swelling noticed by the family. In our case, scrotal swelling was detected by the family. A concomitant Y-chromosomal disorder

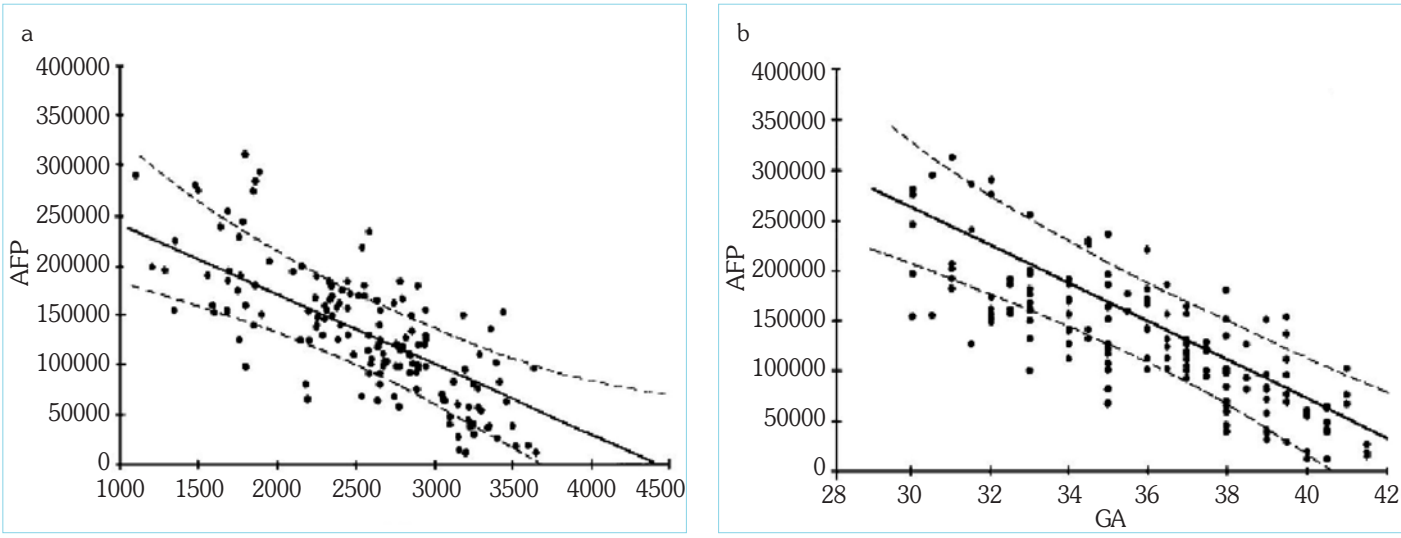

Figure 5. a, b. Changes in alpha-fetoprotein (AFP) levels with body weight (a) and gestational age (b)

Table 1. Cases of juvenile granulosa cell tumor from the literature

Case No	Ref.	n	Right/ Left	Location	AFP level, ng/mL	HCG level, mL U/ mL	Additional pathology	US and size	MRI	Operation	Pathology	Met	Follow-up time
1	Gün et al. (1)	2	I. Left	Scrotum	N	N	-	Complex, multiseptated, hypoechoic, intratesticular solid mass 25×25×20 mm	-	HSO	JGCT	-	-
			II. Right	Scrotum	521.1	N	-	Multicystic, intratesticular, heterogeneous solid mass 25×16×25 mm	-	HSO	JGCT Frozen biopsy: EST/ JGCT	-	-
2	Bulotta et al. (2)	1	Left	Scrotum	6679 IU/mL	8	-	Fetal (36th week) USG: Multicystic testicular mass Postnatal USG: encapsulated, complex cystic mass 15.7×13.3 mm	-	HSO	JGCT	-	5 years
3	Raju et al. (4)	1	?	Intra-abdominal	?	?	XXY (triploidy) ambiguous genitalia	?	?	?	JGCT	?	?
4	Leylek et al. (5)	1	Bil.	Intra-abdominal	?	?	Chromosome 4 deletion, hypotonia, micrognathia, low-set ears, downward-slanted eyes	Very large multicystic, multiseptated intra-abdominal mass	Bilateral large multiseptated cystic abdominal masses	-	JGCT	-	-
5	Peterson et al. (6)	1	Left	Scrotum	55.141	4	-	Complex cystic mass in the left testis-	-	HSO	JGCT	-	
6	Borislav et al. (11)	1	Right	Scrotum	7068	<2	-	Cystic, solid and heterogeneous mass 22×16×15 mm	-	HSO	JGCT (Frozen: GCT)	-	-

Table 1. Cases of juvenile granulosa cell tumor from the literature

7	Levy et al. (12)	6	?	Scrotum	23-61.700	?	?	?	?	?	JGCT	?	?
8	Partalis et al. (13)	1	Left	Intra-abdominal	N	N	-	Cystic mass (cystic intestinal duplication?)	-	Laparotomy 9×9×7 cm	JGCT	-	-
9	Zugor et al. (14)	2	I. Right	Scrotum	35350	N	-	Multiple septic cystic mass (Swiss cheese appearance)	-	HSO	JGCT	-	2 years
			II. Left	Scrotum	9038	N	-	6×6×4 cm Multiple septic cystic mass (Swiss cheese appearance) 5×6×3 cm (Prenatal mass)		HSO	JGCT	-	9 months
10	Yu et al. (15)	1	Bil.	Intra abdominal	?	?	Chromosome 4 abnormality, polymicrogyria renal cysts	?	?	Mass resection	JGCT	-	-
11	Dudani et al. (16)	1	?	Scrotum	?	?	-	-	-	?	JGCT	-	6 months
12	Yıkılmaz et al. (17)	1	Bil.	Intra-abdominal	-	-	Cerebellar dysplasia polymicrogyria	Prenatal (18th week): Normal Postnatal: large multiseptated cystic intra-abdominal mass	MRI: multiple septated cystic mass Right: 8×5×5.5 cm Left: 10.5×7.5×9 cm	Laparotomy, 2 hormone replacement therapy	JGCT	-	-
13	Kos et al. (18)	1	Right	Intra-abdominal	-	-	Cordocentesis: 69 XXY (Triploidy) Inv (9) (p11q13)	Prenatal (26th week): multiple malformations (spina bifida, microcephaly, abdominal tumor) corpus callosum agenesis lung and adrenal hypoplasia	USG: ? multicystic serous fluid containing cysts (5.5×4.1×5 cm) -	?	JGCT	-	-
14	Chan et al. (19)	2	I. Right	Scrotum (3×3×2 cm)	-	-	-	-	-	HSO	JGCT	-	-
			II. Left	Scrotum 18×15×1 mm	27100	-	-	Multicystic mass 3×3×1.5 cm	-	HSO	JGCT	-	-
15	Nistal et al. (20)	1	?	?	?	?	Testicular torsion	?	?	?	JGCT	?	?
16	Uehling et al. (21)	1	Right	Scrotum	450	-	-	Multicystic mass Swiss cheese appearance (Teratoma?)	-	HSO	JGCT	-	-

US: ultrasound; MRI: magnetic resonance image; Bil: bilateral; EST: endodermal sinus tumor; GCT: germ cell tumor; HSO: high scrotal orchiectomy; JGCT: juvenile granulosa cell tumor; Met: metastasis; N: normal; Ref: reference; (-): not specified in article patient details; (?): unreachable patient details

and ambiguous genitalia have been reported in 20% of patients, but we found no Y-chromosome disorder and/or genital anomaly in the present case (4, 5).

Ultrasound can detect an intratesticular multicystic mass, and the normal testicular parenchyma can be evaluated when the clinical diagnosis suggests a cystic teratoma (1, 5-6). In the present case re-

port, we described a complicated cyst with intratesticular septa and indistinct parenchymal contours. This mass lesion was evaluated as a teratoma. As shown in the present case, the MRI signs of this tumor include a multiseptated scrotal mass (1). Although hCG and AFP levels are used as tumor markers, particularly within the first 6 months after disease onset, AFP level do not help in diagnosing a teratoma (7). Bellini et al. (8) demonstrated changes in AFP levels based on body weight and gestational age (Figures 5A and 5B). In such cases, although the AFP level had no diagnostic value during the neonatal period, an increase from a previously low AFP level during the postoperative follow-up period is positively associated with the likelihood of recurrence (9). In our case, the preoperative AFP level was extremely higher than normal, suggesting the diagnosis of a teratoma, but it failed to define the condition. The histopathological examination established the diagnosis of JGCT.

Clinical information about the diagnosis, treatment, and follow-up of JGCTs is primarily in the form of case reports. To date, we found only 24 cases where patients were diagnosed as having JGCTs in the English medical literature (1, 2, 4-6, 11, 12). In total, 6 (25%) of these 24 cases were intra-abdominal, including 3 (50%) bilateral cases. Triploidy (4, 5, 13, 15, 17, 18) a chromosome 4 deletion, and multiple anomalies were reported in the abdominal cases (n=5; 83%). We accessed the data of 12 JGCT cases with the JGCT located in the scrotum; however, no preoperatively diagnosed case of JGCT was found. JGCTs were confused with teratomas, and the diagnosis in all these cases was established based on the histopathology. These were not recurrent cases, and no medical or oncological treatment was applied (Table 1). Therefore, we only performed high scrotal orchiectomy. Organ-sparing surgery has been recommended for small, histopathologically benign tumors with distinct margins. No recurrence or testicular atrophy was reported during the long-term follow-up of these patients (10). We performed orchiectomy because the tumor tissue could not be macroscopically differentiated from the testicular tissue during the surgery.

CONCLUSION

Juvenile granulosa cell tumors are rarely seen, painless, benign masses, particularly in newborns. The preoperative diagnosis of a JGCT is difficult, and it is usually initially diagnosed as a teratoma. Inguinal orchiectomy is a satisfactory treatment modality.

Ethics Committee Approval: Authors declared that the research was conducted according to the principles of the World Medical Association Declaration of Helsinki "Ethical Principles for Medical Research Involving Human Subjects", (amended in October 2013).

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Conceived and designed the experiments or case: TT., ÜB. Performed the experiments or case: TT., ŞC. Analyzed the data: MS. Wrote the paper: TT. All authors have read and approved the final manuscript.

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