

# Juvenile Granulosa Cell Tumor of the Testis: A Bilateral and Synchronous Case. Should Testis-sparing Surgery Be Mandatory?

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Granulosa cell tumor of the testis is an infrequent stromal cell tumor that can be distinguished into adult and juvenile, the latter being more common. Juvenile granulosa cell tumor of the testis is a rare pathologic finding, accounting for 1.2%-3.9% of prepubertal testicular tumors. It is considered as a benign stromal sex cord tumor and is usually unilateral. Although radical surgery was previously considered the treatment of choice, testis-sparing surgery is now recommended in all cases where applicable. We report a bilateral synchronous juvenile granulosa cell tumor in a 6-month-old child treated with testis-sparing surgery and provide a review of the literature. UROLOGY ■: ■—■, 2014. © 2014 Elsevier Inc.

Prepubertal testicular tumors are rare, they account for 1%-2% of all pediatric solid tumors and only 8% are stromal sex cord tumors.<sup>1,2</sup> Juvenile granulosa cell tumor (JGCT) of the testis is a rare pathologic finding, representing just 1.2%-3.9% of prepubertal testicular tumors.<sup>3-5</sup> It is considered a benign stromal sex cord tumor and is usually unilateral. Although radical orchiectomy was previously considered the treatment of choice, possibly because of the poor knowledge of this tumor, testis-sparing surgery can now be recommended and is regarded as the current treatment of choice, where applicable.<sup>4</sup> Furthermore, in last decade, it is well known that the majority of testicular cystic masses in a child are benign lesions or teratomas that can be managed with organ-sparing surgery.

Recently, a case of bilateral JGCT was reported in an 11-year-old boy who was treated with partial orchiectomy and was free of tumor after 6 months of follow-up.<sup>6</sup> In this article, we report the first case of a bilateral and synchronous JGCT in a 6-month-old child who was treated with testis-sparing surgery and provide a review of the literature.

## CASE REPORT

A 6-month-old boy presented with a bilateral palpable masses of the testis after an uncomplicated pregnancy, without prenatal care and with birth at the end of gestation. At birth, the infant was presented with right

cryptorchidism. During a programmed pediatric routine visit, a hard palpable mass of the left testis was detected on physical examination. Testicular ultrasonography revealed a solid and cystic right testicular mass measuring 15 × 10 mm. The left testicle presented with a multilocular cyst of 5 mm in a peripheral area. All images revealed poor vascularization on Doppler study. Abdominal ultrasound was without abnormalities. Testicular tumor markers ( $\alpha$ -FP and  $\beta$ -HCG) were within the normal range:  $\alpha$ -FP, 367.1 ng/mL and  $\beta$ -HCG, <0.100 mIU/mL. Repeat testicular ultrasonography with Doppler (Fig. 1) was performed the day before surgery (3 weeks after the first scan) and showed intraglandular bilateral lesions presenting paths of 12 mm in the lower pole of the right testis and 9.6 mm in the upper pole of the left testis. The lesions displayed similar characteristics, with a solid component and numerous cystic structures representing the predominant pattern. Doppler ultrasound now revealed signs of hypervascularization. Because of the cystic origin of the mass (the solid component was “inside” the cystic lesion), we decided to execute an exploratory inguinoscopy with enucleation of the cystic lesion.

## SURGICAL FINDINGS

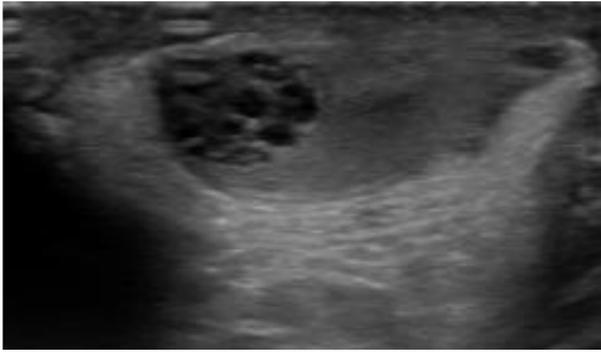
The right testis was occupied by a tumor and a hard mass was present at the upper pole of the left testis. Initially, a left inguinal incision was performed and the left testicular mass was enucleated. A sample from the resection bed was sent to the pathologist. As the specimen proved negative for tumor, the decision was made to perform the same procedure on the contralateral testis. An exploratory inguinal excision on the right side revealed a mass occupying half of the right testis; this mass was

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**Figure 1.** Testicular appearance on Doppler ultrasound examination the day before surgery.

enucleated, similar to the left one (Fig. 2). Although the left testicular remnant was evaluated to represent two-thirds of the total volume of the testis, the right testicular remnant was adjudged to represent only one-half of the testicular volume. An apparently complete tumoral excision was performed. No extemporary section of the tumor was sent to the pathologist according with him. A frozen section of the bed of resection was done and revealed a “normal” parenchyma of the testicle.

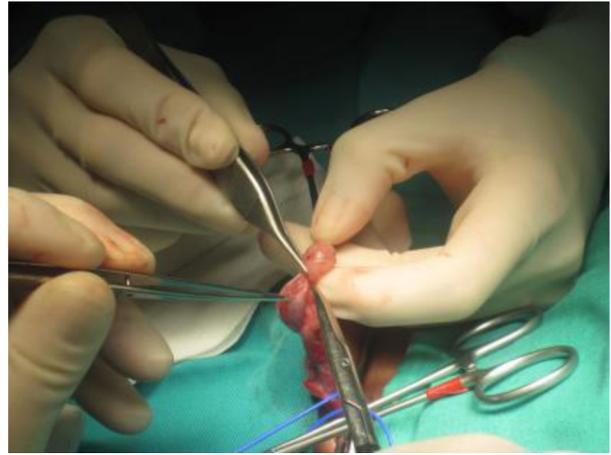
### **PATHOLOGIC STUDY**

The 2 nodular tumors measured 1 cm (right side) and  $0.8 \times 0.5 \times 0.5$  cm (left side) with a smooth external surface and a microcystic appearance in the section surface. Microscopically, both the tumors were cystic and solid. The cystic components are lined for follicular cells with eosinophilic fluid. The solid areas of the cells are arranged in sheets with some myxoid stroma were cystic, with granulosa cells in the wall and basophilic material in the lumen. These cells expressed vimentin and pankeratin. The malignant associated features of sex cord-stromal tumors (size, necrosis, mitosis, and microvascular invasion) were absent in this case and no testicular tumor of this type in testicle has metastasized.<sup>7</sup>

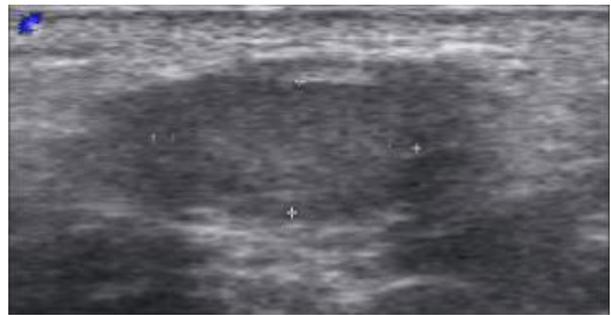
The testicular parenchyma around the tumors was normal. Bilateral juvenile/granulosa cell tumor (JGCT) was diagnosed. Testicular tumor markers were normal at 1 week after surgery. Follow-up ultrasonography (at 4, 8, and 18 months; Fig. 3) revealed that the left testicle was located in the scrotum measuring 12 mm longitudinally. No intraparenchymal lesions were observed. Right testis was in the inguinal canal, measuring 15 mm longitudinally. No intraparenchymal lesions were observed.

### **COMMENT**

Testicular masses in neonates, infants, and prepubertal children are typically benign lesions and most are mature teratomas, epidermoid cysts, or stromal tumors. In these populations, testicular tumors occur with an estimated frequency of 0.5 in 100,000.<sup>5,6,8</sup> JGCT is an uncommon tumor that occurs rarely in the testis. It is usually



**Figure 2.** Appearances at surgery: enucleation of the cystic mass.



**Figure 3.** Follow-up ultrasonography at 12 months after surgery.

described as a single lesion cystic or multiseptated and painless, with normal serum levels of testicular tumor markers.<sup>9</sup> The finding of endocrinological abnormalities in children with testicular lesions should exclude the diagnosis of JGCT,<sup>10</sup> although there has been 1 reported case of JGCT showing hormonal activity.<sup>11</sup> At the same time, the finding of a high level of  $\alpha$ -FP in a 6-month-child should be considered as normal; it is known that its level progressively decreases after birth. JGCTs can be associated with abnormalities of the Y chromosome and ambiguous genitalia. Sometimes, the tumor is diagnosed in the prenatal period<sup>12</sup> and less frequently it presents as an abdominal or inguinal mass in infants with undescended testes.<sup>9,13</sup> Local recurrence or metastases have not been reported in the literature. JGCT usually occurs in the first year of life and the distribution is equal in terms of right vs left.<sup>3,14-17</sup> Although radical orchiectomy was previously described as the treatment of choice,<sup>7</sup> treatment with testis-sparing surgery is now considered safe and is recommended in all cases in which this kind of surgery is applicable.<sup>4-6</sup> The differential diagnosis includes yolk sac tumor, Sertoli cell tumors, teratoma, cystic dysplasia of the testicle, lymphoma, leukemia, and paratesticular masses, such as sarcomas.

During the last 30 years, many studies have evaluated this rare tumor. JGCT is universally considered a benign

lesion with a very good prognosis, but its diagnosis can be made only by the pathologist. Despite this, the increased sensitivity of radiologic imaging studies (which can show multicystic lesions) together with the absence of endocrinological abnormalities can allow this rare tumor to be suspected, and organ-sparing surgery with separate section of the resection bed can then be planned. Such an approach can be considered safe and is now recommended when applicable,<sup>4-6</sup> replacing the previously preferred option of radical orchiectomy.<sup>18</sup> The inguinal approach is globally accepted as the only way for studying any suspicious testicular mass avoiding any different way of access to the testicle. We believe in this kind of surgical approach in the early identification of spermatic chord and vessel control in any case of testicular masses (either malignant or suspicious benign).

As a final point of interest, it is to be noted that JGCT of the testis is histologically similar to JGCT of the ovary and can mimic graafian follicles; furthermore, although it is rare, it is the most frequent congenital testicular neoplasm, representing 6.6% of all prepubertal testicular tumors.<sup>18,19</sup>

## CONCLUSION

To our knowledge, this is the second reported case of a bilateral JGCT presenting as a synchronous testicular mass in a child and treated with testis-sparing surgery. Because of the benign nature of this lesion, testis-sparing surgery should be considered as the treatment of choice in all cases of JGCT, taking into account the surgeon's experience. More generally, we would emphasize that testis-sparing surgery should be used in the treatment of pediatric testicular lesions whenever possible and appropriate.

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