

Case Report

Testicular Sclerosing Sertoli Cell Tumor: Case Report and Literature Revision

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ABSTRACT

Sertoli cell tumors (SCT) are a rare form of presentation of non-germ cell testicular tumors; the sclerosing type is even more infrequent, with less than 50 cases reported in literature. It usually presents in young patients, as unilateral testicular masses.

The aim of this article is to present the case of a 74-year-old male patient, diagnosed with a sclerosing sertoli cell tumor (SSCT), who underwent radical orchiectomy. This type of tumor is a rare variant of testicular neoplasia's; none-the-less, is a differential diagnosis that must be taken into account when there is a small testicular tumor. They usually have a benign and favorable curse, with good prognosis, and do not require any additional management, like in the case of our patient.

INTRODUCTION

SCT are a rare presentation of non-germ cell testicular tumor, and the sclerosing type even more infrequent, with only 50 cases reported in literature [1]. It usually presents in young patients, as unilateral testicular masses, and are benign. Magnetic resonance imaging (MRI) can be a useful tool to diagnose these tumors. They can be managed with radical or partial orchiectomy. The aim of this article is to present the case of a 74-year-old male patient, diagnosed with a SSCT, who underwent radical orchiectomy.

CASE PRESENTATION

74-year-old male patient who first consulted our urology clinic due to left scrotal volume increase, of 6 months of evolution; he denied feeling any testicular masses, nor other symptoms. He had no medical history of importance.

At physical examination, he presented increased left testicular volume, a non-painful left testicle with no masses nor palpable nodules. Testicular tumor markers were negative (alpha-fetoprotein and beta-HGC).

A left testicular ultra-sonogram revealed a solid nodule lesion of 7 mm; and a magnetic resonance showed a nodular lesion of approximately 7 mm, well circumscribed borders, peripheral hypo-intense halo, and heterogeneous hyperintense center in T2 weighted images and isointense in T1 weighted images, that after the administration of contrast had an intense early enhancement (Figure 1).



SCIENTING LITERATURE



SCIENTIFIC LITERATURE



1a: Testicular parenchyma (*RT: right testicle, LT: left testicle*), is slightly hyper-intense. A round, well circumscribed lesion is found in the LT, with a hypo-intense halo and hyper-intense center (arrow). P: penis.

1b: The testicular parenchyma is iso-hypo-intense in these sequences. The lesion is iso-intense to the parenchyma in T1, so it is not visible. With gadolinium, there is a vivid enhancement, complete and homogeneous in the arterial phase that suggests that the lesion is a tumor.

He was presented at the urology medical meeting, and it was decided to perform a radical left orchiectomy, in which we found a hypo trophic left testicle, with a well circumscribed palpable nodule in the back of the testicle. The surgery was performed without complications, and he went home after the procedure.

The pathology report showed a intra testicular tumor lesion of 1 cm of diameter, encapsulated and well circumscribed, that suggested a sex-cord tumor of the sclerosing sertoli type, characterized by sertoli cells with a clear cytoplasm, round nucleus with mild atypia, and central nucleoli, arranged in nests, with tubules and trabeculae, over a collagenous stroma, with no evidence of mitotic activity nor necrosis, with no involvement of the tunica albuginea, tunica vaginalis, rete testis, epididymis nor spermatic chord. Immunohistochemistry confirmed a SSCT, of low grade, with diffuse positivity with vimentin, partial positivity with S100 protein, very focal peripheral positivity with calretinin, and negative with CD117, cytokeratin cocktail, epithelial membrane antigen, inhibin, melan-A, WT-1, CD34, CD31, smooth muscle actin, CD68 and chromogranin; very low Ki67 index (less than 1%) (Figure 2). He has been followed up for 6 months, and no evidence of recurrence of the tumor has been found.



2.1: H-E and positive immunochemistry with vimentin. SSCT. Arrow: tubules. We can observe sertoli cells with a clear cytoplasm, round nucleus with mild atypia, and central nucleoli, arranged in nests, with tubules and trabeculae, over a collagenous stroma, with no evidence of mitotic activity nor necrosis, with no involvement of the tunica albuginea, tunica vaginalis, rete testis, epididymis nor spermatic chord.

2.2: A) H-E, peripheral LT lesion. B) MRI, T2, axial imagen of the LT, at the same level of the H-E image. We can see the excellent correlation between the two diagnostic methods. (Images provided by Dr. Isabel Bolívar, MD, pathologist)

DISCUSSION

SCT are a rare variant of non-germ-cell testicular tumors and represent approximately 1% of testicular neoplasia [2]. There are three types of SCT: general, large-cell and sclerosing [3]. They usually present in post pubertal men ranging from 18 to 80 years [1,2]; they can appear as a unilateral testicular mass with increased scrotal volume and can associate with estrogen release symptoms (usually in younger patients, with gynecomastia in 25% of cases) [1]; they are not related with cryptorchidism [4]. They can be related to Peutz-Jeghers syndrome or Carney's complex [4,5]. Only 10% of SCT are malignant [6].

SSCT were first described in 1992 by Zuckerberg et al [2,7], and since then, only 50 cases have been reported in literature [1], representing only 10% of SCT [6]. The mean age of

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presentation is 35 years-old [8]. They have not been associated with estrogen production released symptoms, and only one case reported by Kao et al, has been reported with metastatic disease [5]. Testicular tumor markers are usually negative [9]. The diagnosis of SSCT is made by histopathology; macroscopically they are well defined nodular lesions, white or yellow [8]. Zuckerberg et al, described the microscopic findings of these tumors: extensive hypo cellular, collagenous stroma separating clusters of sertoli cells, as the case of our patient [5]. The presence of tubular differentiation, and the absence of Reinke crystals helps differentiate the diagnosis of a leydig cell tumor [5].

Immunochemistry has shown the affinity of SSCT for PAX2/PAX8 in nearly 80% of tumors [10], thanks to the extensive fibrosis and the transition process of the mesenchymal

component, even though in testicular tumors this has shown a relationship with lesions from the rete testis and the epididymis. Positive immunostaining with vimentin and inhibin, and negative with keratin and PLAP, associated with the before described microscopically findings are the diagnostic features of SSCT [8], as the ones reported in our patient.

MRI is a powerful diagnostic tool, and can help to differentiate SSCT from other testicular tumors. T2 weighted images are the best for tumor lesions. Tanaka y col, described SSCT as a welldefined nodule, under 15 mm, with generalized hypo-intensity, ssociated with homogeneous enhancement. In T1 weighted images SCT are not well seen or are iso-intense; with gadolinium, there is a quick, avid and homogeneous enhancement [6].The imaging characteristics in MRI studies of SCT are resumed in table 1.

Table 1: Image characteristics in MRI of SCT [4,6].		
	DIAGRAME	IMAGE CHARACTERISTICS
T2 weighted		Best sequence for tumor lesions since it provides an excellent anatomic detail. Tumors are usually homogeneous and hypo-intense (black), or can be iso-intense with a hypo-intense halo (as our case).
T1 weighted		SCT can be iso-intense or not be seen at all. These sequences are sensible to see blood, and exclude other diagnosis as hematomas.
T1 + gadolinium		There is a quick, avid and homogeneous enhancement.

Regarding treatment, due to their benign behavior in 90% of cases, and the usual presentation in younger patients, in most patients a partial orchiectomy is the recommended treatment, only after the diagnosis is confirmed by an intra operative pathological study [8], and when other malignant characteristics have been discarded; these characteristics are: small size (less than 3 cm), high mitotic activity, necrosis, cellular atypia or lympho-vascular invasion [5]. Only with one positive criteria, or when the surgeon believes so, a radical orchiectomy must be performed [4].

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