

Case Report

Sclerosing Sertoli Cell Tumor of the Testis: Case Report and Review of the Literature

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ABSTRACT

Sertoli cell tumors of the testis are extremely rare tumors with a heterogeneous pathology. Three histological variants have been described: Sertoli cell tumor not otherwise specified (NOS), large cell calcifying sertoli cell tumor and the Sclerosing Sertoli cell tumor. The sclerosing Sertoli cell tumor described herein is associated with prominent stromal sclerosis. They present as painless scrotal masses without hormonal disturbances and are benign in nature. Less than 30 cases have been reported in the world literature. We present a case of Sclerosing Sertoli cell tumor in a 39 year old male patient who presented with the complaints of a slowly growing painless mass in the right testicular region for 4 years.

Keywords: Sertoli Cell Tumor, Testis, Case Report, India

Introduction

Sex cord-stromal tumors of the testis are rare and most of them come under the “unclassified” category (1). “Sertoli cell tumors of the testis are extremely rare accounting for 0.4-1.5% of all testicular neoplasms (2)”. These tumors have a heterogeneous pathology (1, 2). Histopathologically three variants have been described: classic (Not otherwise specified), large cell calcifying and sclerosing subtypes (2, 3). These subtypes need to be differentiated as they differ in particular in age of onset, malignant potential, prognosis, and treatment modality

(2). The rare sclerosing variant described herein is a hypocellular tumor with prominent stromal sclerosis and no necrosis or vascular or lymphatic invasion. Dense sclerotic stroma and hypocellularity make the diagnosis on fine needle aspiration difficult as repeated aspirations yield very scanty material. It has a benign behavior and till date, no malignant cases of sclerosing Sertoli cell tumor have been reported. “Both other subtypes have been found to be potentially malignant” (2). Malignant cases have very poor prognosis and it is difficult to treat successfully because of the very little experience available with these tumors (2). Once the Sertoli cell tumor

Received: 05 March 2013

Accepted: 15 July 2013

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has been diagnosed, histological subtyping is necessary as the treatment modality varies. To date, less than 30 cases have been reported (3-12).

We present a case of Sclerosing sertoli cell tumor in a 39 year old male patient who presented with the complaints of a slowly growing painless mass in the right testicular region for 4 years.

Case Report

A 39 year old man presented with a mass in the right testicular region that had been present for 4 years. Tumor markers for testicular cancer such as Beta – Human B and H to capital letters Chorionic Gonadotropin and Alpha Feto Protein levels were not elevated. Repeated fine needle aspiration yielded very scanty material and showed only few spindled cells which no opinion was possible. There was no evidence of estrogen production by the tumor. High orchidectomy was done and the specimen was sent to us for histopathological examination.

Gross examination showed a well circumscribed hard nodular mass measuring 5.5×3.5cm. Cut surface was yellowish white and homogeneous. Normal testicular tissue was not identified (Fig. 1). Microscopy showed sertoli cells arranged in few thin cords, few small tubules and occasional large irregular aggregates in a prominent fibrocollagenous stroma. The tumor cells had a moderate amount of pale cytoplasm and round, bland and uniform nuclei. No invasion was seen. No necrosis and mitosis were seen. The nucleoli were not prominent. The stroma was fibrotic and moderately cellular and hyalinised with few dilated blood vessels (Fig. 2 & 3).

A diagnosis of Sclerosing Sertoli Cell Tumor was made.

Discussion

Sex – cord – gonadal stromal neoplasms of the testes account for approximately 5% of testicular tumors, and most of them are Leydig cell tumors

(2, 12). “Pure Sertoli cell testicular tumors are uncommon and account for approximately 1% of testicular tumors (2)”. “These tumors, usually considered benign, are a heterogenous entity because of their histological and clinical variability” (2,3,12). They are classified according to their histological characteristics into the following subtypes, classic (Not Otherwise Specified), Large cell calcifying variant and sclerosing variant (2, 3, 6). Sclerosing sertoli cell tumor was described by Zukerberg *et al.* in a review of approximately 200 cases of sex – cord – stromal tumors of the testis (1). In their series the sclerosing variant was observed in 10 cases (1). To date, less than 30 cases have been reported (1-12).

The sclerosing sertoli cell tumor is a hypocellular variant with diffuse stromal sclerosis and no necrosis or vascular or lymphatic invasion (1,12). While they can occur at any age, the peak incidence is between 35 and 50 years (1,7,12). In our case the patient was 39 years old. “These tumors are usually asymptomatic, small (0.5 – 2 cm, rarely measuring more than 5.0 cm in their longest diameter), well delimited, white or gray with a solid surface” (1,12). However in our case the tumor measured 5.5 cm in the longest diameter, probably because of the long duration of the swelling. Microscopically, features of Sclerosing Sertoli Cell Tumor include small neoplastic tubules surrounded by a dense sclerotic stroma. These tumors characteristically have small solid cords, anastomosing tubules and thin cords of Sertoli cells in a prominent fibrous sclerosing stroma with thick collagenous bands (1,12). Our case showed sertoli cells arranged in thin cords, few simple tubules, and occasional large irregular aggregates in a prominent fibrocollagenous stroma. “The Sertoli cells are small to medium size with pale cytoplasm and occasional large lipid vacuoles” (1). The nuclei are typically bland. There are usually no mitotic figures but atypia is rarely seen (1). The fibrous stroma may entrap non-neoplastic tubules, resembling immature tubules in Sertoli cell nodules of crypt-

orchid testes (1,12). The tubules may be discrete or anastomosing and may be solid or hollow (1,12). Similar microscopic features were seen in our case also. The sclerosing variant is unilateral and affects either testis at the same rate unlike the large cell calcifying variant which can be bilateral and multifocal (12). "In general, approximately 25% of sertoli cell tumors have estrogen producing activity as gynecomastia, and this feature is absent in the sclerosing variant (12)". Our case also did not show any estrogen producing activity. To date, no recurrent or metastatic cases have been reported (2). The differential diagnosis includes adenomatoid tumor (usually paratesticular, no prominent fibrosis) and metastatic carcinoma (marked atypia). Currently, in the absence of radiologic signs of invasion of lymph nodes or distant metastasis, radical orchidectomy is the recommended treatment.

Here we presented a case of Sclerosing sertoli cell tumor in a 39 year old male patient who presented with the complaints of a slowly growing painless mass in the right testicular region for 4 years. The sclerosing Sertoli cell tumor described above is associated with prominent stromal sclerosis. Although these tumors are usually benign, a long-term follow-up is recommended.

Acknowledgements

The authors declare that there is no conflict of interest.

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