

CASE REPORTS

BENIGN LARGE CELL CALCIFYING SERTOLI CELL TUMOUR OF THE TESTIS: A RARE CASE

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Abstract:

We report a case in which a 14 years old boy with feeling of heaviness of Left testis and also dragging discomfort on the same side with gynecomastia both sides. Ultrasonography showed heterogenous mixed echogenic mass lesion in left hemi scrotum-possibly Germ cell tumor either teratoma or seminoma. His hormonal status was within normal limit except Estradiol which was 150pg/ml. The mass removed by inguinal approach and histopathology confirmed the benign large cell calcifying Sertoli cell tumour (LCCSCT).

The detection of LCCSCTs may indicate to an underlying genetic multiple neoplasia syndrome such as PJS or CNC. Surgery is rarely indicated and aromatase inhibitors constitute an effective treatment for those cases that are associated with gynecomastia and/or advanced skeletal age¹.

Key words: LCCSCT, Carney complex, Peutz-Jeghers syndrome, Gynecomastia.

Bangladesh J. Urol. 2017; 20(1): 46-48

Introduction:

The large cell calcifying Sertoli cell tumour, a distinct sub type, was first described in 1980 by Proppe and Scully. The tumor may occur in an isolated form or may be associated with genetic abnormalities including the Carney complex and Peutz Jeghers syndrome. All of the reported malignant large cell calcifying Sertoli cell tumors are unilateral and unifocal where as 28% benign tumors are bilateral and or multifocal². Tumors that lead to hyper estrogenemia may be treated efficiently with aromatase inhibitors, and change in appearance should prompt evaluation for malignancy.

Case Report:

A 10 years old, student of class-V, hailing from Barisal, presented with—

Left sided testicular swelling for four years which was initially small in size and painless, gradually increasing in size and occasionally associated with a sensation of testicular heaviness with dragging pain in the groin. With

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these complaints he got admitted into the department of urology,SSMCMH.

The apprehensive patient was non anaemic,non-icteric having well developed scrotum with testes of both sides and there was no palpable lymph node elsewhere in the abdomen and groin including supraclavicular region.Patient had bilateral gynecomastia.His left testes was enlarged ,measured about 5cm X 3cm with smooth surface,well defined margins, hard in consistency and non-tender,epididymis was palpable separately and appeared normal.

His scrotal ultrasonography showed heterogenous mixed echogenic mass lesion in left hemi scrotum.Possibly germ cell tumor either teratoma or seminoma and multiple left inguinal enlarged lymph nodes.

His relevant serum tumor markers were assayed.Table-1 showed that all the parameters were within normal limit,except estradiol.

After proper counseling about the operation radical orchiectomy (Lt.) was done through inguinal approach.Cut surface of the mass was homogenous

and calcified structure formed within the testes. The post operative period was uneventful and histopathological examination revealed benign large cell calcifying Sertoli cell tumor.

Table – 1
Report of tumor markers:

Parameters	Patient's Value	Normal value
AFP	0.70 ng/ml	9ng/ml
hCG	1ml U/ml	9mIU/ml
LDH	95 U/L	85-227 U/L
Estradiol	150 pg/ml	12-34 pg/ml



Fig.--1 Showing Gynecomastia

Discussion:

Sertoli cells are the supporting cells of the testicles. The major role of Sertoli cells is to promote the differentiation of spermatocyte. There are four main histological types of Sertoli cell tumors of the testicles: Large cell calcifying (LCCSCT), sclerosing, sex cord with annular tubules and tumors that are not otherwise specified³. Large cell calcifying Sertoli cell tumor is an unusual variant of Sertoli cell tumor.

LCCSCT occur with a frequency of 0.4-1.5% among testicular tumors⁴. They are sporadic in 60% of the reported cases, but in the remaining cases they are linked to multiple neoplastic syndromes such as Peutz Jeghers Syndrome and Carney complex.

Forty nine cases of these tumors have been described in the literature, with patients ranging from 2 to 51 years of age, of the 49 cases, 41 have been reported as benign.

LCCSCTs are first detected in the young from pre pubertal boys to young adults. Due to the frequent higher expression of aromatase within these tumors, there is higher conversion of testosterone to estradiol. As estrogen advances skeletal maturation, LCCSCT in a pre-pubertal boy may present by acceleration of his growth. The continuous effect of estrogens will eventually be noticed as gynecomastia⁶.

Gynecomastia is caused by stimulation of breast tissue from estrogen and is distinct from lipomastia, which is fat tissue around the breast, often seen in overweight boys. With palpation, gynecomastia has the glandular feelings of the breast tissue and unlike lipomastia, the areola can be stimulated. (Fig-1)

Grossly the tumor average 2cm in diameter, are often multifocal, and/are bilateral in about 20% of cases⁷. They are well circumscribed and show a white to tan granular cut surface. Microscopically, the tumor is composed of round to polygonal cells arranged in sheets, nests and trabeculae with central calcification.

Conclusion:

There are strong association of malignant behavior with size >4cm, extratesticular growth, gross or microscopic necrosis, high grade cytological atypia, vascular space invasion, and mitotic rate greater than three mitoses per 10 high power fields. The presence of any one of these features in a solitary large cell calcifying Sertoli cell tumor should be viewed as suspicious for malignant behavior⁸.

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