MIXED SEX-CORD STROMAL TUMOR OF THE TESTIS: A CASE REPORT

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ABSTRACT

Adult testicular granulosa cell tumors are rare sex cord stromal tumors. Mix sex cord stromal tumors are even rare. We report a mixed sex-cord stromal tumor of the testis in which adult granulosa cell component predominates. Case: A 63 year old male patient presented with left hemiscrotal swelling for last 2 years. Physical examination revealed a hard left testicular swelling along with left direct inguinal hernia. Left high inguinal orchidectomy was performed. Microscopy revealed a sex cord stromal tumor. The differentiation was predominantly that of an adult granulosa cell tumor with areas of sex cord stromal tumor with annular tubules and unclassified areas. Only a few mixed sex cord stromal tumor of testis have been reported in literature to determine their biological behaviour. Hence, there is need for long term follow up in these patients.

KEYWORDS: Sex-Cord Stromal Tumor, Granulosa Cell Tumor, Testis

Adult testicular granulosa cell tumors are rare sex cord stromal tumors. Only 31 cases have been previously reported in literature to the best of our knowledge. (Tanner et al;2014) Mixed sex-cord stromal tumors are even rare. We report a case of mixed sex-cord stromal tumor in a 63 year old man with predominant granulosa cell component. Only a few mixed sex cord stromal tumors have been reported in literature to determine their biological behaviour. Hence, there is need for long term follow up in these patients.

Pathological Findings

Left orchidectomy specimen measured 12x10x7cm. Cut section showed a well circumscribed grey white tumor which had entirely replaced the normal testicular parenchyma. (Figure 1) Microscopy revealed a sex cord stromal tumor with a well circumscribed margin. The differentiation was predominantly that of an adult granulosa cell tumor with areas of sex cord stromal tumor with annular tubules and unclassified areas. There was no necrosis and mitotic activity was not prominent. Immunohistochemical staining on paraffin sections showed inhibin positivity in areas of granulosa cell tumor. (Figure 2, 3 and 4) The tumor was vimentin positive (Figure 5) and EMA negative. There was no invasion of tunica, rete, epididymis, spermatic cord or vessels.

DISCUSSION

Sex cordstromal tumors of the testis are uncommon with a spectrum of differentiation including Sertoli-leydig and granulosa theca cell elements. (Allen and Moorehead;1997) The rarest of these tumors is the adult type of granulosa cell tumor and only a few cases have been reported in literature. (Issam et al., 2000) In 1952, Laskowski reported the first case of an adult granulosa cell tumor in a 20-year-old patient. (Laskowski;1952) Most of these tumors are benign, non-functioning and present as painless testicular enlargement or may be detected...
inidentally at autopsy. Some lesions are endocrinologically active presenting with gynaecomastia.

Most adult granulosa cell tumors appear to have benign course, however biological behaviour cannot be accurately predicted from histology alone. These tumors are slow growing neoplasms with potential to form distant metastasis. Histological features that predict malignant behaviour and metastasis include tumor size (>7 cm), vascular and capsular invasion, necrosis and mitotic activity. Recurrences and metastasis can occur late in the clinical course. (Gupta et al; 2008)
Some testicular sex cord-stromal tumors have two or more patterns of histological subtypes and hence are considered mixed. When these tumors are admixed with an unclassified predominant spindle cell component, the term unclassified gonadal stromal tumor is used. Both are grouped together as they have similar clinicopathological features and it is not possible accurately to separate the mixed from the unclassified tumors that have been reported in the literature. (Ro et al; 2002) The immunohistochemical profile of ovarian sex cord stromal tumors has been well characterized, but only few studies have detailed the immunophenotype of the corresponding testicular neoplasms. Inhibin is a peptide hormone that is produced by ovarian granulosa cells and testicular sertoli cells. McCluggage et al found inhibin to be a good marker of testicular sex cord-stromal tumors. (McCluggage et al; 1998) Similar to previous studies, the tumor exhibited strong positive staining for inhibin and vimentin in the present case.

Management of sex cord-stromal testicular tumors is still debated, since there are insufficient number of these cases reported in literature to determine whether they follow any particular behaviour pattern.

Rarity of such tumors makes it difficult to develop a consensus on a standardized approach and individual lesions are perhaps best assessed according to their predominant component of differentiation.

To conclude, Only a few mixed sex cord stromal tumors have been reported in literature to determine their biological behaviour. Rarity of these tumors makes it difficult to develop a consensus on a standardized approach of management and individual lesions are perhaps best assessed according to their predominant component of differentiation. Hence, long time follow up of these tumors is warranted.