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A Rare Encounter of Intra-Abdominal Seminoma with Testicular Dysgenesis Syndrome

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Abstract Undescended testis is a known risk factor for testicular cancer and intra-abdominal malignant changes are not uncommon. We present a case of a 34-year-old male who had an intra-abdominal classical testicular seminoma, bilateral cryptochidism and hypospadias. Patient has all the features of "Testicular dysgenesis syndrome". This is an increasingly common problem in male reproductive health. In most cases, TDS presents with impaired spermatogenesis, but the more severe spectrum will include genital malformations and testicular cancer. Current scenario is unexpectedly found at surgery for cryptorchidism or inguinal hernia, while imaging play a vital role for findings of cited condition for adequate preoperative planning to avoid damage of fertile testes and vasa deferens.

Keywords Abdominal Malignant; Testicular Seminoma; Testicular Dysgenesis Syndrome

1. Introduction

Testicular dysgenesis syndrome (testicular feminization syndrome), is a clinical spectrum which includes a high incidence of cryptorchidism and hypospadias, low semen quality and an increasing incidence of testicular cancer.[1] Progressively trend was observed in literature especially in the western countries.[2]

Testicular dysgenesis leads to inhibition of spermatogenesis and supremacy of sertoli cells (feminizing), also known as an androgen insensitivity syndrome (AIS).[3] Thus, such patients despite having a male genotype present as asymptomatic, functionally normal but reproductively sterile females. AIS is caused by mutations in the androgen receptor gene and is associated with abnormal testicular development with augment threat of germ cell malignancy.[4] The risk of neoplasia is also recognized in a maldescended testis and often increases with advance ages.[5]

2. Case Report

A 34 years old single male, presented to the Urology clinic complaining of a bloated abdomen for the last 4 months. Patient has discomfort sensation rather than sharp pain, also complained of lethargy, loss of appetite and loss of weight (4 kg over the past 4 months). Upon examination a right iliac fossa mass extending slightly to the lumbar region was noted; it was immobile, non-tender firm and measured 6cm x 4cm. As shown in figures 1 and 2, clinical examination revealed abnormal genitalia with a mal-developed scrotal sac, bilaterally cryptorchidism, and an underdeveloped penis with a peno-scrotal hypospadias. Patient had the problem since birth but never seek treatment so far.

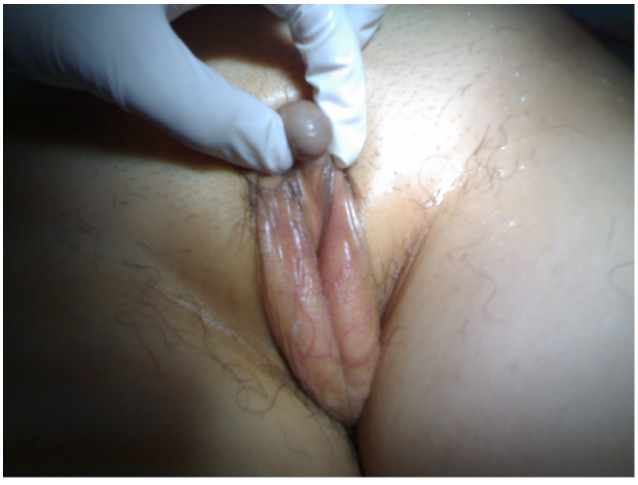

Figure 1. peno-scrotal hypospadias

Computed tomography revealed an intra-abdominal mass which suggested malignancy of an undescended testis in the abdominal cavity most likely representing a seminoma. Furthermore, left testis was not found. Patient serum beta-human chorionic gonadotropin was high but alpha-fetoprotein levels and lactate dehydrogenase levels

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were normal. Further hormonal studies showed an increase in Luiteinizing Hormone (LH) and Follicle Stimulating Hormone (FSH) but a very low Testosterone level which pointed to primary testicular failure. Patient proceeds to laparotomy and the tumor was removed en-bloc (figures 3 and 4).

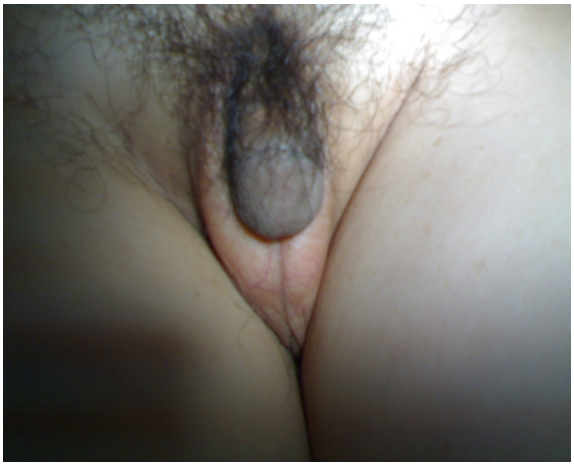

Figure 2. underdeveloped scrotum and penis

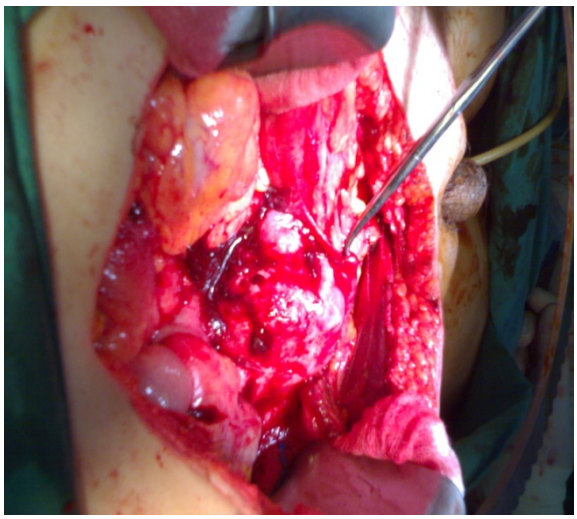

Figure 3. intra-operative findings of the seminoma

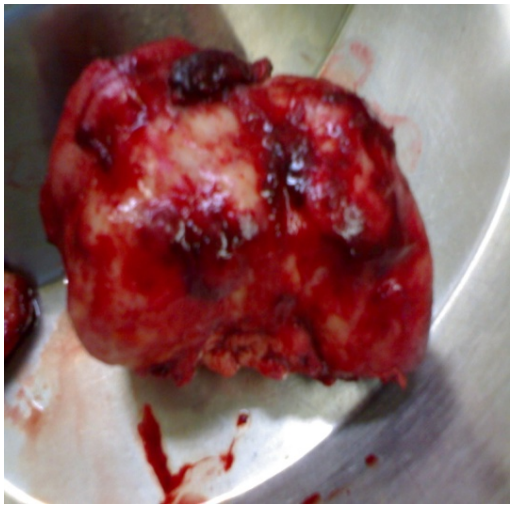

Figure 4. Tumour mass- seminoma

Histologic examination showed the mass to be a classical Seminoma. Patient was subsequently given radiotherapy and jointly followed-up in the oncology clinic and urology clinic.

3. Discussion

There is well-established evidence for increased risk for testicular cancer in maldescent testis and intra-abdominal tumor.[6] The clinical expression of symptoms in testicular dysgenesis syndrome may vary, even within a syndrome caused by a single gene defect. The most severe forms of testicular dysgenesis syndrome, e.g. in individuals with 45X/46,XY karyotype (and a high percentage of aneuploid cells), often include three or four symptoms, including undescended testis, impairment of spermatogenesis, hypospadias and/or testicular neoplasia. Noverthless, patients with a less severe form may have one or perhaps two symptoms.[1] Whereas, It is a rare finding for an adult male to present with an undescended testis, which is usually detected during routine postnatal assessment.[1]

The accepted hypothesis is that early adverse (intrauterine) environmental factors and genetic polymorphism lead to impaired Leydig and Sertoli cell differentiation subsequently androgen insufficiency and impaired germ-cell differentiation eventually lead to Testicular Dysgenesis syndrome. Presentations vary from mere subfertility to patients with 3 or 4 symptoms like present patient illustrated above.[7]

The Leydig cells differentiate and produce testosterone to induce masculinization whereas Sertoli cells organize themselves into testicular cords surrounded by peritubular myoid cells and enclose fetal germ cells. Rare genetic disorder can also contribute to syndrome (for example: 45X/46XY and androgen insensitivity) but it's not common and 80% of the patient will have no genetic disorder.[7] Testicular cancer mostly arises from CIS (carcinoma *in situ*) which are postulated to have originated from germ cells that escape normal differentiation *in utero*. Several epidemiological studies support this view.[8]

Rising data from clinical observations of individual patients indicate incline trend of male reproductive problems, like genital abnormalities, reduced semen quality, sub-fertility and testicular cancer. Geographical relations and multiple problems in one individual, strongly suggest the existence of a pathogenetic link. The relationship of male reproductive problems is perhaps not coincidental but shows the existence of a common cause resulting in a maldeveloped testis. Still there is a doubt that the testicular dysgenesis syndrome can be a result of disruption of embryonal programming and gonadal development during fetal life.[1]

4. Conclusions

It is a known fact that un-descended testis is a risk factor for testicular seminoma; however in practices the dealing

with incidental findings of abdominal mass with absent unilataral testis and presence of hypospadias, care about 'Testicular dysgenesis syndrome. Suspected patient should be further investigated by hormonal readings.

REFERENCES

[1] Skakkebaek NE, Rajpert-De Meyts E, Main KM. Testicular dysgenesis syndrome: an increasingly common developmental disorder with environmental aspects. Hum Reprod 2001; 16:972–8.

[2] Devine and Horton, 1977. Devine Jr. CJ, Horton CE: Hypospadias repair. *J Urol* 1977; 118:188-193.

[3] Rutgers J. L. and Scully, R. E. "The androgen insensitivity syndrome (testicular feminization) a clinicopathologic study of 43 cases," International Journal of Gynecological Pathology 1991; 10 (2): 126–144,

[4] Hannema, S. E. Scott, I. S. Rajpert-De Meyts E., Skakkebæk, N. E. Coleman, N. and Hughes, I. A. "Testicular development in the complete androgen insensitivity syndrome," Journal of Pathology, 2006; 208 (4): 518–527.

[5] Karabulut, N. Karabulut, A. Pakdemirli, E. Sabir, N. Soysal, S. K. and Soysal, M. E. "Stromal tumor of the sex cord in a woman with testicular feminization syndrome: imaging features," American Journal of Roentgenology, 2002; 178 (6) : 1496–1498.

[6] Giwercman, A., Bruun, E., Frimodt-Moller, C. and Skakkebaek, N.E. (1989) Prevalence of carcinoma in situ and other histopathological abnormalities in testis of men with a history of cryptorchidism. *J. Urol.* 142, 998-1001.

[7] Griffin, 1992. Griffin JE: Androgen resistance—the clinical and molecular spectrum. *N Engl J Med* 1992; 326: 611-618.

[8] CristiÃ¡n Palma C; CristÃ³bal B; Maccioni R, New case of an intra-abdominal testicular seminoma in an adult, Actas Urol Esp, FEB-2007; 31(2): 160-3.