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Bilateral Cryptorchid Malignancy with Persistent Mullerian Duct

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Introduction

Testicular carcinoma is the most common solid tumor in males between 35 and 39 years; however, it has been reported before 10 and after 50 years, as well.[1] Persistent mullerian duct syndrome (PMDS) is a rare form of male pseudohermaphrodism secondary to absence or deficiency of anti-mullerian hormone (AMH). This hormone which is produced by sertoli cells during embryonic stage suppresses mullerian ducts and impedes the formation and development of female internal genital organs.[2] Genetically, this factor can be transmitted heterogeneously and in various forms. Short arm of chromosome 19 is the place of a gene which produces AMH.[2,3]

Case Report

A 23-year-old man was referred to our hospital because of undescended testis (UDT) and abdominal mass. Physical examination revealed empty hypoplastic scrotum. No testis was palpable in scrotum and inguinal canal. A palpable and movable mass was palpated on abdominal midline. Other physical examinations were normal. The patient was married and had infertility and azooespermia. External genitalia and sexual function was normal. Two pelvic masses and a lesion at the behind of bladder were reported in pelvic ultrasonography (fig. 1). Serum tumor markers were measured; FP was higher than normal and HCG was undetectable. For further study, abdominal and pelvic CT scan were done in which the same reported masses by ultrasonography and retroperitoneal lymphadenopathy were reported, as well. The patient underwent laparotomy with a probable diagnosis of UDT. During surgery bilateral large tumor of testicular cancer in UDT was observed together with mullerian remnants including uterus, fallopian tube and cervix extended to upper part of prostate (fig. 2). No ovary was seen in pelvic exploration. Testis tumor and mullerian elements were removed, the pathology of which indicated bilateral classic seminoma with embryonal carcinoma associated with uterus, fallopian tube, and cervix (fig. 3).

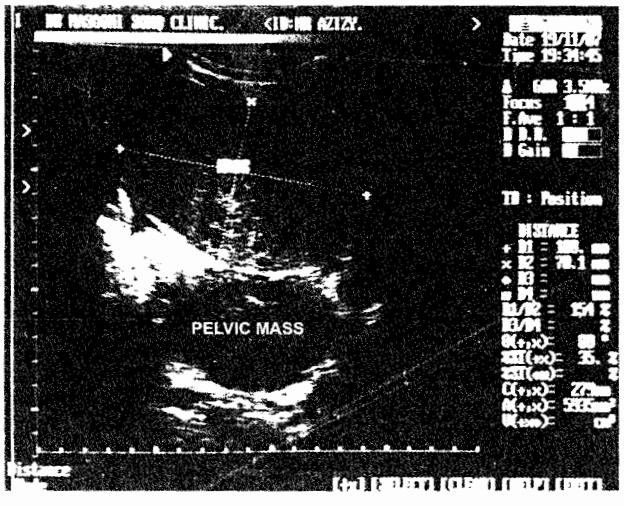

FIG. 1. Pelvic masses and a lesion at the behind of bladder were reported in pelvic ultrasonography.

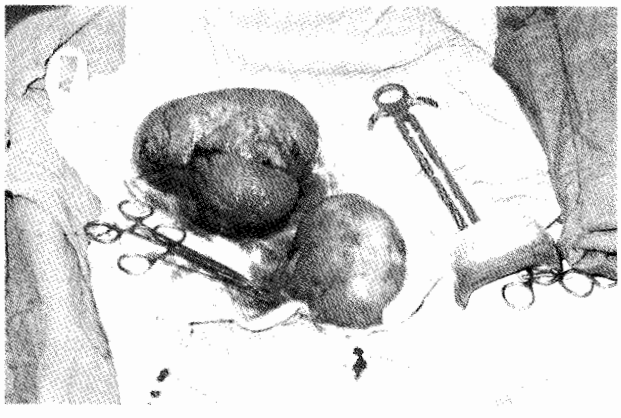

FIG. 2. Bilateral large tumor of testicular cancer in UDT together with mullerian remnants including uterus, fallopian tube and cervix extended to upper part of prostate

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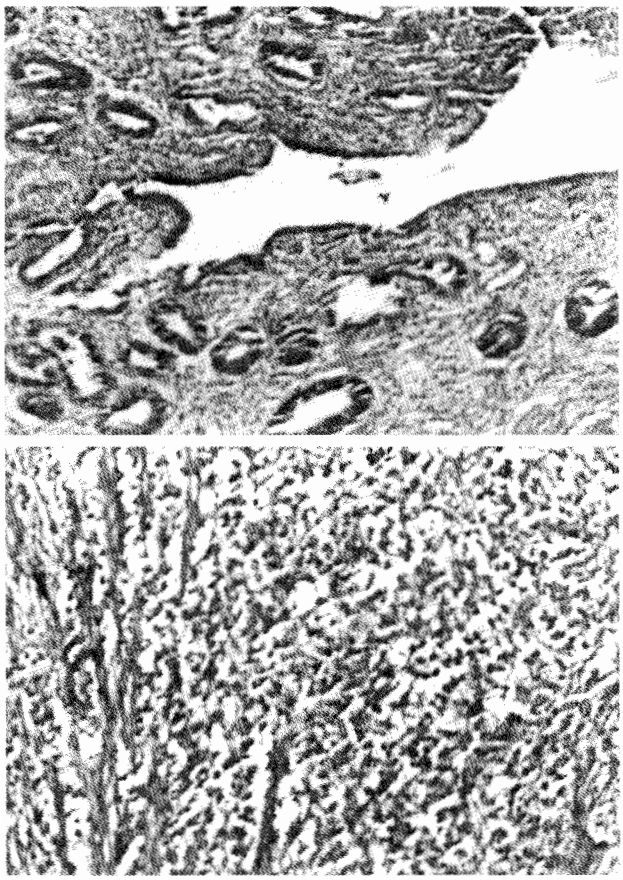

FIG. 3. Bilateral classic seminoma with embryonal carcinoma

Discussion

The most common types of testicular tumors are the ones with germ cell origin (90-95%), of which seminoma and mixed germ cell tumor are most common types.[1,4,5] Within the category of mixed cell types, the majority (up to 25% of all testicular tumors) are teratocarcinoma which are a combination of teratoma and embryonal carcinoma.[1,4,5]

In our respective case, reported pathology was sominoma and embryonal carcinoma. Cryptorchidism is one of the major etiologic causes for testicular cancer, as 7% to 10% of patients with testicular tumors have a history of UDT.

Trauma, hormonal disorders, and testicular atrophy are among acquired risk factors accounting for testicular tumor. It is obvious that all acquired factors do not lead to testicular tumor formation; however, they can be considered as a parameter in detecting testicular cancer (such as trauma). There is no particular tumor marker for typical seminoma. Serum HCG only increase in 5 to 10% of cases.

The combination of increased serum FP and seminoma indicates the existence of non-seminomatous elements in testicular tumors.[1,6] Searched in Medline, just one case of testicular tumor associated with the absence of AMH in which, mullerian duct derivatives included uterus, fallopian tube and cervix together with abdominal unilateral testicular seminoma was reported.[2]

In this case mullerian remnants were found next to the tumoral testes, which were classic bilateral seminoma and emberional carcinoma (fig. 3).

Seminoma and NSGCT (non-seminomatous germ cell tumor) in UDT has been reported in several articles;[4,5] however, the association of bilateral tumor and mullerian derivative has not been yet reported and only one case of unilateral tumoral UDT with mullerian elements has been reported. Some believe that the absence of AMH does not increase the incidence of testicular tumors, but others believe that this deficiency can increase the mentioned incidence by 5 to 15%.[4] Thus, it is not an indication for orchidectomy. These testes can lead to normal UDT with absence of AMH. Routine orchidectomy would not be recommended and the existence of testes can lead to virilization and only orchopexy must be done. Vas deferens should not be injured to protect fertility.

In the cases of complete removal of female internal organs, vas deferens may be injured; hence, cervix and the upper third of vagina should be left if exists.[1]

Since in this study both testes were tumoral, bilateral orchidectomy with complete removal of mullerian remnants were performed and the patient was referred for radiotherapy.

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