Para-testicular florid cystic müllerianosis in an elderly patient: Case report of a rare metaplastic change of tunica vaginalis with unusual clinical presentation (mass forming lesion)

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**ABSTRACT**

Müllerian metaplasia/florid cystic Müllerianosis of the tunica vaginalis is one of the metaplastic changes described in the testis. Others include urothelial metaplasia, epidermoid metaplasia, and gastric metaplasia. Florid cystic Müllerianosis shares similar histo-morphological features and immunohistochemical profile with female counterpart endosalpingiosis. Herein, we report a case of para-testicular florid cystic Müllerianosis presented as a painless testicular mass in a 66-year-old male with normal tumor markers.

1. Introduction

Florid cystic Müllerianosis of the testis is a recently described entity. The histogenesis of these lesions is not clearly understood. Some authors postulated an origin from a Müllerian embryonic remnant, while others considered it as metaplastic process of the lining mesothelial cells of the tunica vaginalis. Histologically, the lesion is made up of variable-sized cysts. The lining epithelium resembles that of fallopian tube epithelium with ciliated columnar cells, non-ciliated secretory cells, and ‘peg’ cells. Immunophenotypically, these lesions express progesterone receptors (PR) in the epithelial cells and the stroma, while CD10 is expressed in the surrounding connective tissue stroma, reminiscent to female endosalpingiosis profile. An extensive literature search revealed only three reported cases of Müllerian metaplasia of testis/para-testis. Herein, we describe a case of a gentleman who presented to the Urology clinic with a painless testicular mass with an initial clinical consideration of testicular malignancy. The patient underwent an uneventful right radical orchietomy. The microscopic examination and immunohistochemistry markers support the diagnosis of Müllerian metaplasia/Florid cystic Müllerianosis in para-testis. Herein, we report a very unique and an exceedingly rare case of Testicular Müllerian metaplasia/Florid cystic Müllerianosis.

2. Case presentation

A 66-year-old male patient known case of hypertension complained of on-and-off bilateral testicular discomfort for more than eight years. He had no fever, pain, hematuria, or urinary difficulty. The patient has no family history of malignancy. MRI was suggestive of a right testicular tumor, 6 mm in maximum dimension, (Fig. 1), with no evidence of any other organ abnormalities and no evidence of metastasis. Physical Examination of the external genitalia demonstrated a hardness of the right testis; though non-tender with a negative transillumination test. The left testis was normal, homogenous with grade II varicocele on the Valsalva maneuver. No clinical evidence of gynecomastia nor cryptorchid testicles. Laboratory investigations revealed normal hormonal serum levels including alfa-fetoprotein and beta-HCG serum levels. Urine culture was negative. He underwent right radical orchietomy. The right testis weighted 48.0 g (6.5 × 4 × 2 cm), and the spermatic cord (10 × 1.2 cm). Cut sections of the testis demonstrated a solitary, firm, well-circumscribed, yellow mass (0.5 × 0.5 × 0.5 cm), primarily arising from and abutting the tunica vaginalis. However, the remaining testicular parenchyma, epididymis, and spermatic cord were grossly unremarkable.

Histologically, the area of concern was small (5 mm in the greatest dimension), comprised of variable-sized glands within the tunica vaginalis; some are cystically dilated with evidence of histiocytic reaction and cholesterol clefts suggestive of a reaction to rupture. They are lined...
by simple flat to cuboidal epithelium comprised of three types of cells: ciliated columnar cells, Non-ciliated secretory cells, and ‘peg’ cells, (Fig. 2A and B. The surrounding stroma is a non-endometrial type. The remaining testicular parenchyma, seminiferous tubules, and the interstitium were unremarkable with no evidence of cytological atypia, necrosis, or mitosis. The glands were immunoreactive for PAX-8, WT-1, CAM 5.2, and CK8/18 while negative for SALL-4 and CD117 (Fig. 3A and B). The postoperative period went uneventful, and the patient is doing well on follow-up.

### 3. Discussion

There are four types of metaplasia described in the literature to arise from the testicular tunica vaginalis; gastric metaplasia, epidermoid metaplasia, urothelial metaplasia, and Müllerian metaplasia\(^1\). Urothelial metaplasia being the most common type, with a proportion of 17%, Sundarasivarao.\(^2\) To the best of our knowledge, Müllerian metaplasia is exceedingly rare, only three reported cases in the Literature by Nistal M. Microscopically, all cases show aggregate or rosary bead arrangement of cysts, lined by cuboidal to pseudostratified epithelium comprised of tubal-like cells: ciliated, secretory non-ciliated, and ‘peg’ cells and surrounded by a connective tissue stroma; indeed, the aforementioned histological features are similar to our present case. PAX-8, WT-1, CK8/18, and CAM5.2 were positive. Altogether suggests a metaplastic Müllerian derivative epithelium.

Given the high frequency of malignancy in mass-forming lesions of the testis, the clinical decision was to go for a radical orchiectomy. In the present case, no evidence of germ cell neoplasia. The immunoprofile is negative for CD117 and SALL-4. Various theories postulate that the histopathogenesis of Müllerian metaplasia/Florid cystic Müllerianosis occurs either as a metaplastic change of the mesothelial cells in the tunica vaginalis\(^4\) or as an embryonic Müllerianosis remnant in...
men confined to glands and small ductus of the spermatic cord and prostatic utriculus. Although florid cystic Müllerianosis has a benign behavior, previous studies documented a malignant transformation in rare cases into ovarian-like serous carcinoma of para-testicular structures (serous cystadenocarcinoma). 3-5

4. Conclusion

We report a very unique and an exceedingly rare case of Testicular Müllerian metaplasia/Florid cystic Müllerianosis. Awareness of metaplastic changes that might involve testis and/or para-testis is essential to reach a precise diagnosis, especially when dealing with mass-forming lesions, to avoid misinterpretation and misdiagnosis. The florid cystic Müllerianosis characterized by both morphological and immunoprofile that resembles the female counterpart endosalpingiosis.

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