CASE REPORT

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Papillary Cystadenoma: Unusual Localization

Özgen ARSLAN SOLMAZ[a]

[a]Clinic of Pathology,
Elazığ Training and Research Hospital,
Elazığ

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Correspondence:
Özgen ARSLAN SOLMAZ
Elazığ Training and Research Hospital,
Clinic of Pathology,
Elazığ, TURKEY
ozgensolmaz_73@hotmail.com

ABSTRACT Papillary cystadenoma of the epididymis is a rare epithelial tumour which is thought to develop within the efferent ductules. Only 60 histologically documented cases have been reported in the English-language literature since the original report by Sherrick in 1956. A 61 years old man presented to urology department complaining of a pain in his left scrotum. Enlargement of the left scrotal had been noticed 1 year ago. Macroscopically, the tumor was grey white in colour, cystic appearange and measured 3x3x2,6 cm in size. Microscopy revealed a cyst cavity, containing papillary structures supported. Because of papillary cystadenoma of the epididymis was reported that transformation to cystadenocarcinoma and it can be seen with the von Hippel-Lindau disease, This risk should not be forgotten in the epididymal cysts and surgical treatment should be done in this direction.

Keywords: Epididymis; cystadenoma, papillary; von hippel-lindau disease

Papillary cystadenoma of the epididymis (PCE) is the second commonest benign neoplasm of this organ following adenomatoid tumor.[1] Its is a rare epithelial tumour which is thought to develop within the efferent ductules.[2] It was first described in 1956.[3] It may occur sporadically or as a manifestation of von Hippel-Lindau disease (VHLD).[4] Only 60 histologically documented cases have been reported in the English-language literature since the original report by Sherrick in 1956.[5]

Because of papillary cystadenoma of the epididymis is a rare epithelial tumour and was reported that transformation to cystadenocarcinoma and it can be seen with the von Hippel-Lindau disease, we wanted to present this case.

CASE REPORT

A 61 years old man presented to urology department complaining of a pain in his left scrotum. Enlargement of the left scrotum had been noticed 1 year ago. On examination he was found to have painless epididymal masses. Left epididymal mass was 3x3 cm in size and was firm, mobile and non-tender. The radiological images of the patient could not be reached. Under general anesthesia, a 3 cm well-circumscribed tumor was excised along with the left epididymis. Left epididymal mass was removed and sent for histopathological examination.

Macroscopically, the tumor was grey white in colour and measured 3x3x2,6 cm in size. Cut surface showed multiple cysts filled with yellowish

serous fluid. Microscopy revealed a cyst cavity, containing papillary structures supported. This was lined by cuboidal cells, some of which were ciliated. The cyst lining was in continuity with a rim of residual epididymis. Immunohistochemical profile showed positivity for PAX-2, Vimentin, Cytokeratin AE1/AE3, CD10 negativity for CEA, CD20, EMA, S100, RCC (Figure 1). The diagnosis of papillary cystadenoma was made based on morphological and immunohistochemical features.

DISCUSSION

Tumors of the epididymis comprise only 5% of intrascrotal neoplasms and most are benign. Papillary cystadenoma of the epididymis is the second most common benign tumour following adenomatoid tumour. Other benign tumours of epididymis include leiomyoma, lipoma, melanotic neuroectodermal tumour of infancy, mixed gonadal stromal tumour and cavernous haemangioma.[1]

In the majority of cases, PCE develops within the efferent ductules of the head of the epididymis as a partially or completely cystic or solid lesion measuring about 1–3 cm in diameter and is usually asymptomatic.[2-6] However, in symptomatic patients the most common presentation is a painless, slowly growing, scrotal swelling. In rare occasions, they may present with pain or tenderness in the scrotum or are found to have an epididymal nodule during an examination for infertility. The ages of occurrence are 16–65 with a mean age of 36. Most common location is the head of epididymis. It may occur sporadically or as a manifestation of von-Hippel Lindau disease.[7,8]

PCE of the epididymis may involve one epididymis or both epididymides. According to Odrzywolski, unilateral PCE has never been reported as the initial presentation of VHLD. Bilateral PCE occurs mostly in patients with previously diagnosed VHLD, but in some cases there is no evidence of other stigmata of the disease.[9]

Papillary cystadenoma of the epididymis is typically less than 5 cm in size, although the reported size ranges from 0.5 to 8.0 cm (mean, 2.1 cm; median, 2.0 cm).[10] In this case, mass is 3 cm. Similar to this case, grossly, PCE forms a well-circumscribed or encapsulated, variably cystic nodule, which may be tan or yellow and histologically, is characterized by cysts filled with prominent intracystic papillary projections. Cell borders are prominent. Cilia are occasionally present. The supporting stroma may be vascular, collagenous (in our case too), or inflamed. Nuclear atypia, stratification, mitoses, necrosis, psammoma bodies, and invasion of surrounding structures are absent.[9]

Metastatic renal cell carcinoma may appear to be histologically similar to papillary cystadenoma and both may be seen in patients with VHL. Immunohistochemically, the epithelial cells of PCE are consistently positive for cytokeratin AE1/AE3, CAM 5.2, and epithelial membrane antigen (EMA). The keratin profile is CK7 positive and CK20 negative. Renal cell carcinoma marker, carcinoembryonic antigen, vimentin, and S100

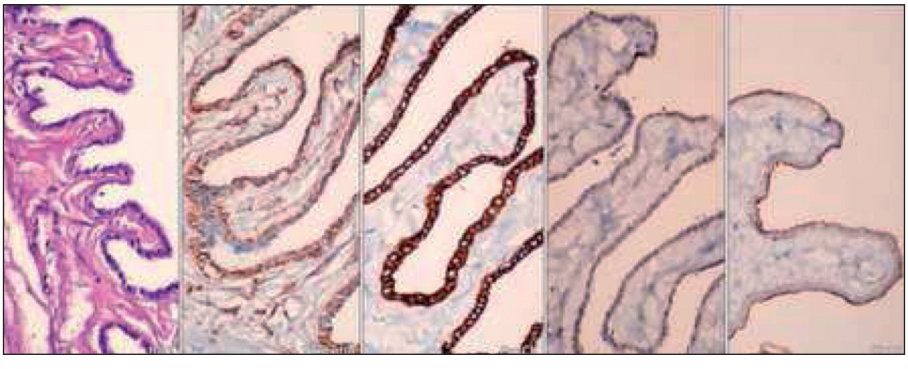

FIGURE 1: Collagenous papillary structure (Hemotoxylen eosin x200) and positive staining with Vimentine, Pancytokeratin, PAX-2 and CD10 (Immunochemistry x200).

are variably positive. PAX2 has been reported to be positive in one case. CD10 has been negative in all cases tested. Positivity for CK7 and negativity for CD10 were recently successfully used to demonstrate a rare case of a tumor-to-tumor metastasis from a clear cell RCC to a PCE.[9] In this presented case immunohistochemical profile showed positivity for PAX-2, Vimentin, Cytokeratin AE1/AE3, CD10 negativity for CEA, CD20, EMA, S100, RCC.

Treatment of PCE consists of testicle-sparing surgical excision. Local excision is usually feasible, although some patients do undergo orchiectomy. It has been suggested that patients be followed after excision. There is one report of recurrence (possibly due to incomplete initial excision) and 2 reports of transformation to cystadenocarcinoma.[11]

We describe the morphologic features and immunohistochemical staining pattern of one case of papillary cystadenoma in epididymis in the case of VHDL which the PCE is usually bilateral. In unilateral cases of PCE such as our patient's, literature reviews provide evidence against extensive and expensive genetic testing for VHLD. Because of PCE was reported that transformation to cystadenocarcinoma and it can be seen with the von Hippel-Lindau disease, This risk should not be forgotten in the epididymal cysts and surgical treatment should be done in this direction.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

This study is entirely author's own work and no other author contribution.

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