Serous papillar y adenocarcinoma of the rete testis: Unusual ultrasonography and pathological findings

ABSTRACT
We report a case of serous papillary adenocarcinoma of the rete testis in a 22-year-old man. Adenocarcinoma of the rete testis is highly resistant to radiotherapy and any known chemotherapeutic regimen. We recommend radical orchietomy. At last follow up, the patient was well, without any evidence of recurrence, ten years after surgery.

KEY WORDS: Rete testis, serous papillary adenocarcinoma, orchietomy

INTRODUCTION
Adenocarcinoma of the rete testis is a rare neoplasm of the male genitourinary tract. It usually occurs in men after the age of 60 and carries a variable prognosis. Less than 30 cases of Mullerian-type serous tumors occurring in male patients were reported. Adenocarcinoma of the rete testis is highly resistant to adjuvant radiotherapy and any known chemotherapeutic regimen with activity against ovarian malignancies. We report the case of a young man with a malignant serous papillary adenocarcinoma of the rete testis achieved a complete remission and remained free of disease after ten years.

CASE REPORT
A 22-year-old man was referred to our institution for the evaluation of a left scrotal mass which developed within six weeks after being treated four weeks by antibiotics for alleged epididymitis with no response. The patient complained of pain, swelling in the right scrotum and subfebrile temperature. He was investigated by ultrasonography. CT scan was negative for retroperitoneal lymph nodes. The serum level of CA 125 was normal for female values. As missed torsion of the testicle was suspected, exploration via a scrotal incision was performed. The intraoperative findings were: enthickened tunica vaginalis, medium quantity of yellowish fluid in the virtual cavity between the two layers of the tunica vaginalis, gray nodular mass between the lower pole of the testis and the epididymis. Radical orchietomy were performed. After obtaining results of pathological findings, a right hemiscrotectomy was performed. No further treatment was given. The patient is alive with no evidence of disease for ten years since surgery.

Ultrasonography findings
Testicular ultrasonography confirmed the presence of a marked multiseptated accumulation of fluid. The unusual finding delineated on this side was the nodular and irregular anterior thickening of the paratesticular structures [Figure 1a and 1b].

The testis was normal in size and echostructure. The accumulation of fluid and the nodular thickening of the paratesticular envelope of the testis

Figure 1: (a) Irregular thickening of paratesticular tissues anteriorly (arrows). (b) Severe septated hydrocele
raised suspicion of mesothelioma with malignant potential. Transvers ultrasonography of the left testis after evacuation of hydrocele and open biopsy of the paratesticular tissues was performed. There is severe thickening of the extratesticular layers anteriorly. The epididymis is not distinguishable. Irregular cystic and solid spaces and hyperechoic punctate structures [Figure 2] representing psammoma bodies are demonstrated (the unusual prominent features of the tumor).

The head of epididymis was not distinguishable from the peripheral thickening. Color doppler investigation of the mass showed a hypervascular pattern. Hyperemia is a well-recognized feature of scrotal inflammatory disease. However neoplastic disease is also commonly hypervascular especially in larger lesions.[1]

**Histological findings**

Clusters of papillary tumor located in parapenidimal soft tissue, invading tunica vaginalis were found. Most papillae lined by cuboidal cells, small part, by columnar, non ciliated cells. Tumor cells showed malignant features with enlarged pleomorphic nuclei and nucleoli. Few mitotic figures were detected. Micropseudocyst formation and intracystic papillae were present. Detached small solid clusters infiltrating stroma, were also seen. Numerous psammoma bodies were detected, in association with tumor cells and in stroma [Figure 3].

Neutral mucin stain (mucicarmin) showed positive mucin drops in tumor cytoplasm. Immunohistochemical results showed intense positivity for CEA, Keratin, EMA (membranous and cytoplasmic) CA-125 and Vimentin LUM1 was positive in only few tumor cells.

The distinction of serous papillary carcinoma from malignant mesothelioma was the main diagnostic problem. The diagnosis of serous papillary carcinoma in this case was based on the prominent papillary growth pattern, numerous psammoma bodies, positive immunostaining, mainly for CEA and CA-125, the patient young age and the lack of anamnestic asbestos exposure.

**DISCUSSION**

Less then 30 cases of papillary adenocarcinoma of the rete testis were published. In the differential diagnosis, one should consider paratesticular tumor arising from the mesothelial surface of the tunica vaginalis[2] and malignant mesothelioma. However, as neither the slit-like tubules characteristic of carcinoma of the rete testis nor transition between epithelial cells of the normal rete testis and the neoplastic cells could be identified. In our case there was no history of asbestos exposure, which is sometimes present in patients with mesotheliomas. Psammoma bodies, which were prominently seen in this case, are more common in serous papillary adenocarcinomas than in mesotheliomas. These tumors involve either the visceral tunica vaginalis and underlying tunica albuginea or, more commonly, the upper pole of the testis, particularly in the region of the testiculoepididymal groove.[4] Clinically, paratesticular serous papillary carcinoma occurs in young to middle-aged adults and typically presents as a testicular mass, which may be associated with a hydrocele but only five cases associated with hydrocele were described in the literature. Our case is the first case of multiseptated accumulation of fluid ultrasonographically diagnosed. Neoplastic disease, paticular larger lesions, are commonly hypervascular. The tiny unusual calcifications within the mass were consistent with psammoma bodies found in the pathological specimen. Serous papillary adenocarcinoma of the rete testis is with malignant potential and metastasis in less than one year.[5] Only in one case, a seven-year interval was described between initial presentation and development of clinically metastatic disease.[4] Our case had no signs of recurrence ten years after surgery, left radical orchiectomy and hemiscrotectomy.

**Figure 2:** Irregular cystic and solid spaces and hyperechoic punctate structures (arrows) representing psammoma bodies.

**Figure 3:** Papillary tumor cluster, surrounded by reactive fibrous tissue and psammomatous bodies (right lower corner), H and E, x400.
The treatment of choice is orchiectomy because highly resistant to adjuvant radiotherapy and any known chemotherapeutic regimen with activity against this malignancies.

REFERENCES


Source of Support: Nil, Conflict of Interest: None declared.

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