Case - Ovarian-type cancer in males: Paratesticular serous papillary carcinoma

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INTRODUCTION

Paratesticular serous papillary carcinoma (PSPC) is a rare testicular tumour analogous to ovarian epithelial cancer, with approximately 50 reported cases since 1966. It originates from Müllerian (paramesonephric) duct remnants in males, particularly the appendix testis, retained in up to 83% of adult males.[1] Due to its rarity, the natural history and optimal management remain poorly understood. We present the case of an 84-year-old male diagnosed with metastatic PSPC.

Case report

An 84-year-old man presented with progressive dyspnoea after two months of malaise. Previously independent and with good exercise tolerance, he now required oxygen and was found to have bilateral pleural effusions. CT showed multiple pulmonary nodules.
The patient reported a right testicular mass since adolescence. A sonogram 10 years before admission showed a normal right testicle with a moderate hydrocoele, calcification, and tubular ectasia of the left rete testis (figure 1).

Four years prior to admission the patient had right testicular discomfort. Repeat imaging showed the hydrocoele remained moderate, with a 3 mm right epididymal head cyst and a hypoechoic focus extending into the inguinal canal, initially suspected to be a hernia (figure 2). CT confirmed the cyst and calcification but no hernia.

Two months prior to admission, follow-up ultrasound revealed a vascular, lobulated hyperechoic mass arising from the right epididymis or tunica vaginalis (figure 3). Differential diagnoses included adenomatoid tumour, lipoma, leiomyoma, and sperm granuloma. On exam, the mass was fixed and appeared to invade the overlying scrotal skin.

The patient was referred for radical orchiectomy, but his comorbidities and onset of acute hypoxic respiratory failure precluded surgery. Bedside percutaneous biopsy was performed. Histology showed papillary structures with moderate pleomorphism and psammoma bodies (figure 4). Immunohistochemistry supported a diagnosis of PSPC with tumour cells demonstrating strong diffuse positivity for cytokeratin 7 (CK7), Ber-EP4, Wilms tumour protein 1 (WT1), estrogen receptor (ER), and paired box gene 8 (PAX-8), and with wild-type p53. In the paratesticular region, mesothelioma or some metastatic carcinoma (such as primaries from lung, GI tract, thyroid or testicular germ cell tumors) can be other differentials as they also can present with papillary formation on histology. However, mesothelioma or metastatic carcinoma have been excluded as the tumor cells do not stain for CK20, CK5/6, TTF1, thyroglobulin, CDX2, NKX3.1, calretinin, D2-40, GATA3, CEA, AFP, CA19-9, CD15, CD10 and CD30.
CT revealed multiple pulmonary lesions and bilateral effusions without adenopathy or ascites. Overall suggesting metastatic spread of the scrotal mass to the lungs in the absence of another primary malignancy, for instance breast which could also be ER positive.

Subsequent consultation showed invasion of adjacent stroma and further confirming the diagnosis of paratesticular low-grade serous papillary carcinoma.

Pleural fluid cytology was positive for malignancy, with papillary structures and psammomatous calcification. Immunohistochemistry matched the paratesticular tumour with aspirated cells positive for CK7 and Ber-EP4, confirming metastatic spread.

The patient was referred to oncology for systemic therapy, but due to poor functional status, palliative care was pursued. He died shortly thereafter.

DISCUSSION

Although histologic parallels between ovarian and testicular germ cell tumours are well established, ovarian-type epithelial tumours in the testis are exceedingly rare.[2,3,4] The WHO classifies testicular tumours as germ cell tumours, sex cord–stromal tumours, and miscellaneous tumours, the latter including Müllerian-type epithelial neoplasms.

The embryologic basis for PSPC lies in the incomplete regression of the Müllerian ducts in males. Embryonic development is sexually indifferent until the sixth week of gestation, at which point the mesonephric ducts become the male genital duct system and the paramesonephric ducts become the female genital duct system. In males, anti-Müllerian hormone-driven involution of the paramesonephric ducts leaves behind Müllerian vestiges known as the appendix testis and utricle.[5,6] The entire regression of the Müllerian system, which in females develops into the uterovaginal primordium and salpinges, occurs in one day.[5] The appendix testis is present in up to 83% of testes.[1]

Incomplete regression results in vestigial structures like the appendix testis; the site from which PSPC can arise. Its histology and immunophenotype mirror those of serous carcinomas of the ovary.

Due to its rarity, PSPC is primarily described in case reports. Most reports describe longstanding scrotal masses or hydrocoeles with a small solid component and calcification, features seen in this case.[7,8,9,10] Several cases note that imaging initially suggested benignity, often leading to delayed diagnosis.[3,11,12]

This case underscores the importance of re-evaluating long-standing hydrocoeles when new features appear, such as calcification or solid components. The indolent course of PSPC can mask malignant potential until metastases arise.

Management is typically surgical, with radical orchiectomy as the mainstay. In cases with metastatic spread, chemotherapy modelled on ovarian cancer protocols has been attempted. There is no established standard of care due to the condition's rarity.

CONCLUSIONS

PSPC is a rare malignancy often masquerading as benign hydrocoele. Its indolent nature and nonspecific features pose diagnostic challenges. This case highlights the need for vigilance when imaging reveals atypical features in scrotal masses. Given its origin from Müllerian remnants, PSPC represents a male counterpart to ovarian serous carcinoma and warrants greater awareness among clinicians.

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FIGURES AND TABLES

Figure 1. Sonogram 10 years before admission with right-sided hydrocoele.

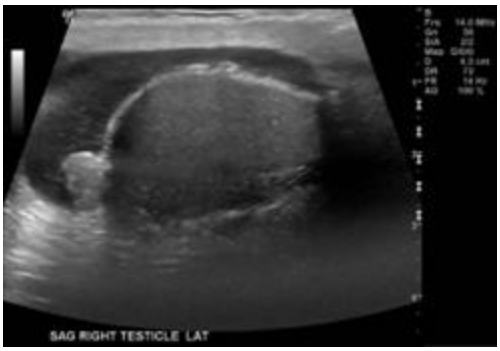

Figure 2. Sonogram 4 years before admission with epididymal cyst and echogenic debris.

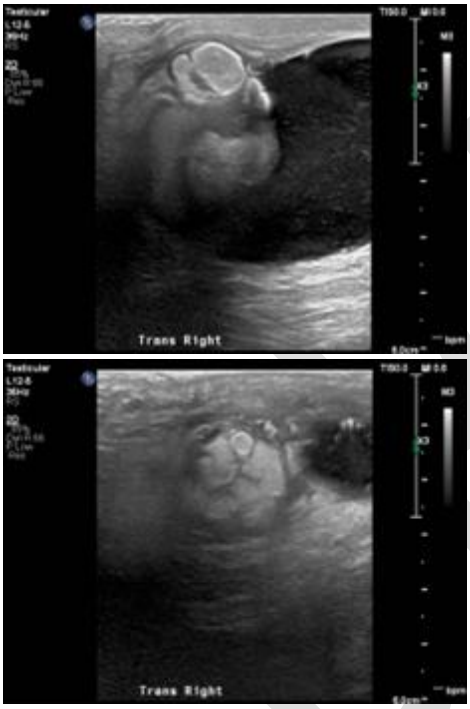

Figure 3. Sonogram 2 months before admission with a vascular lobulated mass in the right hemiscrotum.

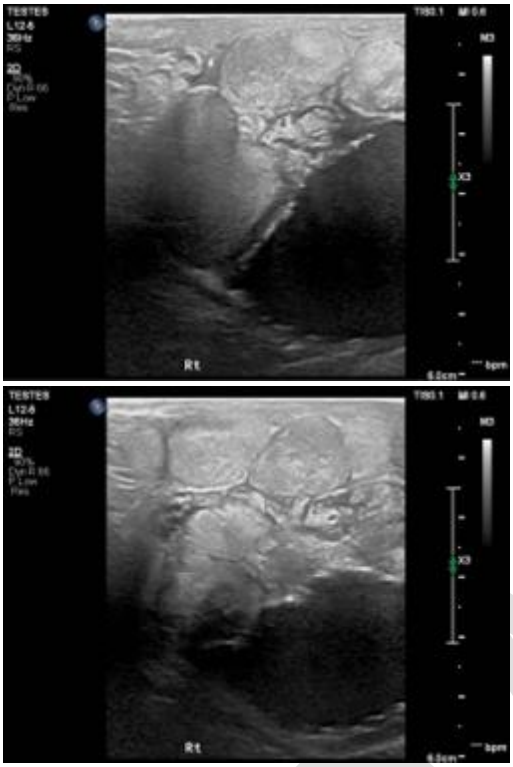

Figure 4. Biopsy from the right paratesticular lesion. (A) low magnification view; (B) high magnification. The biopsy of the right paratesticular mass is composed of micro-papillae that are lined by a simple cuboidal epithelium with low-grade atypia. There are very rare mitoses present. Haemorrhage and necrosis are not appreciated. The micro-papillae are embedded in a sclerotic stroma and numerous psammoma bodies are seen (psammoma bodies, yellow arrow). At the edge of the cores, invasion of the adjacent stroma is seen (invasive tumor, red arrow).