Extragonadal mixed germ cell tumor of the seminal vesicle

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ABSTRACT
Primary malignancies of the seminal vesicles are rare. Presentation of extragonadal germ cell tumor in the seminal vesicle is even rarer. We report a case of a 26-year-old male who presented with hematuria and lower urinary tract symptoms, which on imaging turned out to be a right seminal vesicle mass. The diagnosis of mixed germ cell tumor with yolk sac tumor and teratoma was made on the initial evaluation by transrectal ultrasound-guided biopsy which showed the characteristic histomorphology and immunohistochemistry profile. The patient underwent chemotherapy followed by radical pelvic exenteration. The patient is doing well with no evidence of distant metastasis in positron emission tomography/computed tomography of 1-year posttreatment.

INTRODUCTION
Extragonadal tumors of the seminal vesicle are a very rare disease, with only a few cases reported in the literature.1,2 We present a 26-year-old male with nonseminomatous extragonadal mixed germ cell tumor arising from the seminal vesicle as the primary site with no evidence of tumor in the testis or retroperitoneum.

CASE REPORT
A 26-year-old male presented with complaints of hematuria and lower urinary tract symptoms. On digital rectal examination, a hard mass was felt in the pelvis, whereas there was no palpable mass per abdomen or in the testes. Both the testes were felt normally descended in the scrotum. No imaging revealed any suspicious pathology in the testis. Computed tomography scan (CT) revealed a mass arising from the right seminal vesicle and prostatic region measuring 6 cm × 6.7 cm × 7.6 cm3. Magnetic resonance imaging of the pelvis confirmed the findings further. Transrectal ultrasound (TRUS)-guided biopsy was performed. Microscopic examination showed the features of mixed germ cell tumor with yolk sac tumor, teratomatous, and glial tissue components. Immunohistochemistry showed strong positivity for CK (AE1/AE33), vimentin, alpha-fetoprotein (AFP), SALL4, glypican-3, and CD117 [Figure 1a-j]. Serum tumor markers were elevated [Table 1]. Chest CT showed no focal lesion. On initial evaluation before chemotherapy induction, the patient had a mildly deranged liver function test (LFT), which was attributed to antibiotic therapy by the hepatologist. The patient was started on (etoposide [EP] and cisplatin) chemotherapy regimen for the first cycle. After 2 weeks, the patient’s LFT became normal and the chemotherapy was stepped up to (bleomycin, EP [BEP], and cisplatin) regimen. After completion of the second cycle, the patient presented with passage of urine through anus and pneumaturia, suggestive of rectovesical fistula. The patient underwent a diversion colostomy. Later, chemotherapy was...
completed with a total of one cycle of EP and three cycles of BEP regimen. Tumor markers showed a significant decline in the levels postchemotherapy [Table 1].

Postchemo positron emission tomography/CT revealed tumor activity in the right seminal vesicle region. Further, CT scan showed a large mass in the right seminal vesicle measuring 6 cm × 6.7 cm × 7.6 cm³ invading almost all the surrounding structures including the left seminal vesicle, prostate, bladder, and rectum with associated rectovesical fistula [Figure 2]. In view of the above findings, the patient underwent radical pelvic exenteration surgery. Gross examination of the specimen showed a blackish pigmented lesion in the right seminal vesicle infiltrating the prostate, bladder, and rectum [Figure 3]. Histopathology confirmed tumor origin being the right seminal vesicle with infiltration of the rectal wall, urinary bladder wall, and prostatic stroma. Predominance of teratoma with high-grade glioma-like areas with glial overgrowth exhibiting nuclear atypia, necrosis, and microvascular proliferation. No immature neuroepithelium was identified. Immunohistochemistry showed diffuse strong expression for glial fibrillary acidic protein (GFAP) with Ki67 proliferation index of 5%–1%. The tumor showed no expression for AFP. Pelvic lymph nodes were negative for metastasis.

**DISCUSSION**

Malignancies of the seminal vesicle are rare. Usual primary malignancies seen in the seminal vesicle are carcinomas or sarcomas. In our patient, TRUS-guided biopsy of the seminal vesicle mass revealed malignant mixed germ cell tumor with yolk sac tumor and teratoma. Immunohistochemistry confirmed the presence of yolk sac tumor, which showed positivity for SALL4, AFP, and glypican-3. Extragonadal germ cell tumors are seen usually in the retroperitoneum or mediastinum. We confirmed our findings from careful physical examination and scrotal ultrasound to rule out any testicular primary. The diagnosis on needle biopsy enabled planning for appropriate treatment protocol.

One interesting observation is the presence of high-grade glioma-like areas which showed strong expression for GFAP.

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**Table 1: Serum tumor markers**

| Tumor markers | Reference values | Before starting chemo | After two cycles of chemo | After four cycles of chemo |
|---------------|------------------|-----------------------|--------------------------|--------------------------|
| Beta HCG      | Male <5 mIU/ml   | 262.2                 | 5.2                      | 0.9                      |
| AFP           | 85% of population <8.5 ng/ml | 3176 | 51 | 20.4 |
| LDH           | <200 U/L         | 526                   | 178                      | 202                      |

AFP = Alpha-fetoprotein, LDH = Lactate dehydrogenase, HCG = Human chorionic gonadotrophin
Teratomas can undergo malignant/somatic differentiation in approximately 6% to 14% of cases. In a study by Matoso et al., neuroglial differentiation and neoplasms in 13 testicular germ cell tumors were analyzed. The study showed the absence of ATRX, IDH, or BRAF alterations in the tumors, indicating different oncogenic mechanisms than their CNS counterpart. Four patients received chemotherapy with BEP.

The most appropriate treatment for neuroglial neoplasms arising in teratoma is unclear due to insufficient data. Since teratomas are resistant to chemoradiotherapy, surgical resection is the gold standard treatment. The patient after 1 year of follow-up after surgery is currently without evidence of disease.

**CONCLUSION**

Extragonadal mixed germ cell tumor arising from the seminal vesicle with no evidence of disease in the testis, mediastinum, or retroperitoneum is uncommon. It may be managed successfully with chemotherapy followed by surgery.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.