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Background

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract, frequently involved the stomach, small intestine, and rectum. Extra-GIST (E-GISTs) are similar in histology and immunohistochemistry to GIST but arise outside the gastrointestinal tract. E-GISTs represent only 5-10% of all GIST cases, but primary prostatic GISTs are extremely rare. Only a few cases were reported in the literature.^{1,2}

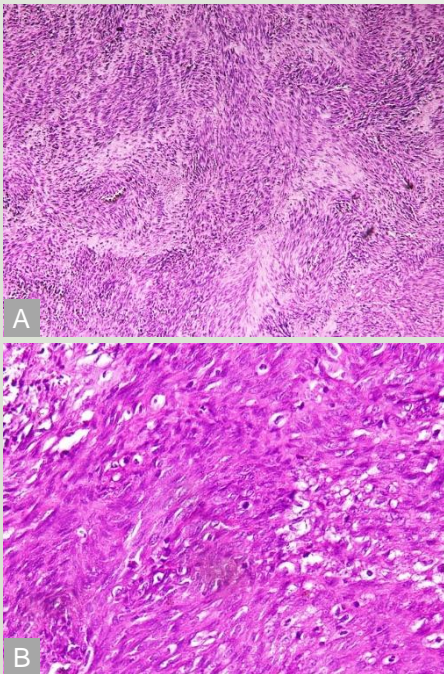


Figure 1. Most of tissue fragments were colonized by cellular spindle cell neoplastic proliferation with a fascicular growth pattern (A). The neoplastic spindle cells had a moderate degree of atypia with eosinophilic cytoplasm and indistinct cells borders and mitotic count of >5/50 HPF (B) (HE, 100x, 400x).

Case Description

A 63 years-old male presented urine retention in the last week. Dysuria, hesitancy, urinary frequency, and nocturia were felt in the past sixth months and got worse. He had no history of hematuria, urethral catheterization, or urinary tract stones. Physical examination was unremarkable and systemic examination was essentially normal. Digital rectal examination found a markedly enlarged prostate gland with a smooth and bulging surface, firm consistency and nontender. Hematology tests and urinalysis was normal. Prostate-specific antigen (PSA) level was 0.11 ng/ml (cut off \leq 4 ng/ml). The chest X-ray indicated normal lung and heart. An enlarged prostate with 11.46 cm in diameter from ultrasonographic. The abdominal CT scan showed a large lobulated mass in pelvic cavity. It was poorly defined in some part, with multiple calcifications that pushed up the urinary bladder and rectum wall. It was pointed to prostate. It also showed hydronephrosis bilateral and multiple lesions in right liver. The scan excluded the presence of another gastrointestinal or extra-gastrointestinal primary tumor.

A gross examination from transurethral resection of prostate (TURP) revealed pieces of white tissue with brown parts, elastic-solid, measuring 2.5 x 2.5 x 2.5 cm. The specimen was entirely submitted for microscopic examination. Histologically, hematoxylin and eosin-stained sections showed that most of tissue fragments were colonized by cellular spindle cell neoplastic proliferation with a fascicular growth pattern. The neoplastic spindle cells had a moderate degree of atypia with eosinophilic cytoplasm, indistinct cells borders and mitotic count of >5 per 50 high-power fields (HPFs) (Figure 1). There was some necrotic mass with calcification. Immunohistochemically, the tumor was positive expression for CD117 (c-kit), CD34, and DOG-1 while smooth muscle actin (SMA) and S-100 were negative (Figure 2). The final diagnosis consistent with a malignant GIST of prostate.

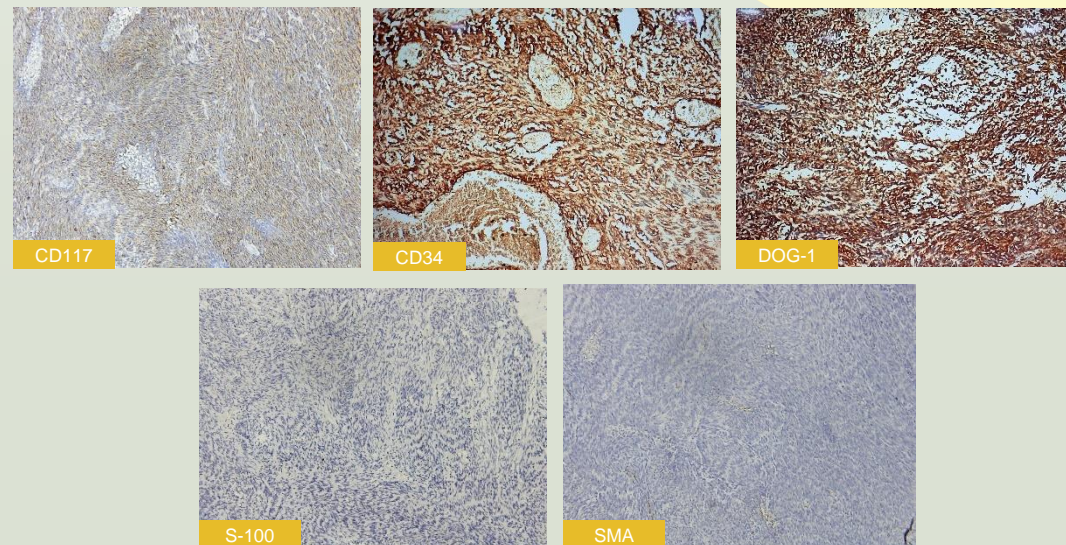


Figure 2. The tumor was positive expression for CD117 (c-kit), CD34, and DOG-1 while S-100 and SMA were negative (100x).

Discussion

The first case of prostatic E-GIST was reported by Van Der Aa et al. in 2005.^{1,3} As far as we know, only 18 cases have been reported in English literature. Herein, this present case should be the 19th literally reported primary prostatic E-GISTs.

Due to the lack of cells of Cajal in the E-GIST site, some pathogenesis theories proposed that it can arise from ectopic interstitial cells of Cajal. These cells originated from the pluripotential progenitor of mesenchymal cells.¹

In the diagnosis of E-GIST, clinician should first remove the possibility of a tumor originating from the gastrointestinal tract.⁴ Clinical and radiological examination play crucial role in rolling out of possibility of rectal GIST secondarily invading and involving the prostate. Al-Maghrabi et al. reported 90% cases demonstrate a prostatic mass compressing the rectum and urinary tract system but restricted to the organ's capsule.¹

The concept of tumor risk assessment is crucial for potential assessment of biological GISTs. The criteria risk stratification includes tumor size and mitosis per 5 mm² or 50 HPF. Tumors with a diameter <5 cm are typically at low risk, while >5 cm are malignant. Mitotic rates >5 per 50 HPFs usually characterize GISTs as malignant.^{1,3}

Immunohistochemistry plays a great role to confirm the final diagnosis of GIST. The positive ratio for CD117, CD34, and DOG-1 of E-GISTs were about >81%, 50% and 92%, respectively.^{3,4}

Conclusions

It is crucial to distinguish prostatic E-GIST from other mesenchymal spindle cell tumors. The combination of imaging, histological and immunohistochemical play a critical in substantially diagnosing a primary E-GIST, especially when arising from an unusual location. Expression of CD117, CD34, and DOG-1 confirmed the diagnosis.

References

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