Case Report

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Case Report: Very Rarely Synovial Sarcoma with an Intrapelvic Location

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Abstract

Computed lower abdomen tomography and pelvic magnetic resonance imaging were performed in a 59-year-old male patient who presented with complaints of urinary retention and pelvic pain. It was observed that there was a mass lesion with a heterogeneous internal structure, located in the midline in the pelvic region, with a lobulated contour, approximately 14x13 cm in size, pushing the bladder forward and the rectum backward, containing cystic and contrasting solid areas. The patient underwent pelvic exploration and mass excision. These recent pathological findings were consistent with the diagnosis of synovial sarcoma (SS). SSs are tumors that occur mostly in the para-articular soft tissue of the extremities in young adults and are extremely rare in the primary pelvic region.
Keywords: Intrapelvic mass, synovial sarcoma, acute urinary retention

Introduction

Synovial sarcoma (SS) constitutes approximately 5 to 10% of all soft tissue sarcomas. It is a high-grade spindle cell tumor with t (X; 18) (p11;q11) chromosomal translocation detected in more than 95% of cases (1,2). SS is frequently detected in the extremities, especially in the periarticular region, and rarely shows intra-articular localization (3,4). Clinically, they appear as a palpable and painful soft tissue mass. It can also occur in other body parts such as the neck, tongue, larynx, mediastinum, the esophagus, heart, lung, abdominal wall, small bowel mesentery, vessels, and retroperitoneum. The intrapelvic location of the SS has been reported very rarely in the literature. Our aim in this case; to contribute to the literature by examining intrapelvic SS, which is a place where SS is very rare, clinically, radiologically, and histopathologically.

Case Report

A 59-year-old male patient was admitted with complaints of acute urinary retention and pain in the pelvic region. On digital rectal examination; fairly large, soft mass was palpated. The serum prostate-specific antigen level was measured at 1.31 ng/mL. In the complete urinalysis, eight erythrocytes were seen in each field and the urine culture was sterile.

A giant solid mass lesion in the pelvic region and bilateral hydroureteronephrosis due to possible mass compression were detected the abdominopelvic computed tomography (CT). In the pelvic magnetic resonance imaging (MRI) examination performed with an intravenous contrast material for the localization and characterization of the mass; a giant mass lesion with a heterogeneous internal structure was observed, located in the midline in the pelvic region, with a lobulated contour, approximately 14x13 cm in size, pushing the bladder anteriorly and the rectum posteriorly, containing cystic and contrasting solid areas. At the level of the pelvic floor, it was observed that the mass pushed the prostate of normal size forward, resulting in the displacement of the prostate toward the symphysis pubis. It was noted that the mass was separate from the prostate but closely adjacent to the posterior prostate capsule. Extraprostatic pelvic mesenchymal tumor or tumor originating from the prostate capsule was considered in the radiological differential diagnosis (Figure 1).

After the examination of the patient, his preoperative preparations were completed, his informed and informed

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consent was obtained, and he was hospitalized with a pelvic exploration planned. Under general anesthesia, in the supine position, after providing appropriate sterility conditions, a suprapubic transverse skin incision was made. The bladder was deperitonized. The bladder was opened with the help of a scalpel. In intraoperative observation; in the prostatic lodge after opening the bladder; the prostate was of normal size on inspection. However, upon the observation of giant cystic mass that pushed the bladder anteriorly, originating between the prostate and rectum and extending superiorly to the level of the umbilicus, the incision area was enlarged median below the umbilicus. A 5 cm long incision was made on the wall of the cystic mass. Approximately one liter of hemorrhagic cyst content and necrotic soft tissue pieces were removed from the cystic mass. Frozen tissue samples were sent for the diagnosis of pathological tissue. The frozen report; the tumor that is very rich in small cells, came as a mass that required examination of paraffin sections for malignancy exclusion. After the bleeding control was achieved, the layers were closed in the appropriate plan and the operation was terminated.

The tissues taken intraoperatively were sent to the pathology laboratory for pathological examination (Figure 2). It was decided to differentiate between benign and malignant tumors paraffin sections, as the diagnosis of frozen tissue was reported as a tumor rich in small cells by microscopic examination of irregular tissue pieces, which were macroscopically observed as 6 cm in diameter, and were sent for frozen examination. In the macroscopic examination of the material, which was completely sent after the operation, 14x13x1 cm, yellow-white colored, soft consistency, mostly necrotic tissue pieces were observed, which were considered irregular tissue pieces. On microscopic examination, monotonous tumor cells with small and spindle morphology, hyperchromatic nuclei, narrow cytoplasm, and a spindle pattern distribution within areas of intense necrosis and fresh bleeding were noted (1,5). No prostate and rectal tissue were observed in the histopathological examination of the tumoral mass, which was clinically expressed as originating between the prostate and rectum. In the immunohistochemical examination; while no staining was observed with pan-cytokeratin, desmin, smooth muscle actin, CD34, CD31, c-kit, and DOG-1, diffuse, strong, cytoplasmic staining was observed with vimentin. Approximately 50% proliferative activity was observe with Ki-67, and the case was reported as SS due to extensive, strong, cytoplasmic staining with TLE and BCL-2 (Figure 3) (1,2,5).

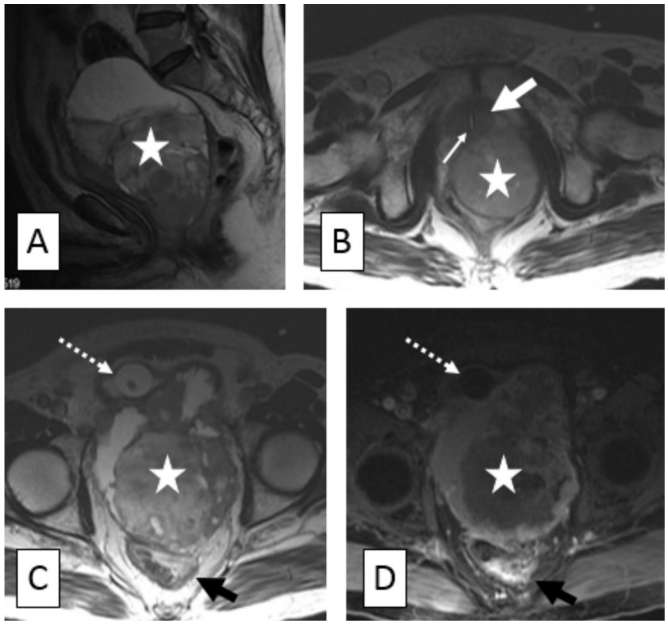

Figure 1. MRI examination; A. Sagittal T2 weighted image, B. Transverse T2 weighted image at the the pelvic floor, C. Transvers T2 weighted image, D. Transverse contrast-enhanced T1 weighted image.

A giant pelvic mass with cystic and solid areas (star) located between the bladder (dashed white arrow: urethral catheter balloon in the bladder lumen) and rectum (black arrow). The mass is adjacent to the prostate and pushes the prostate (white arrow) forward (thin white arrow: catheter in the prostatic urethra lumen)

MRI: Magnetic resonance imaging

Discussion

A SSs is are tumors originating from undifferentiated mesenchymal tissue and constitutes 5-10% of all malignant mesenchymal tumors. It shows slow growth in an expansile character. Usually 3-5. occurs between decades (2). The male/female ratio has been reported as 2/3. Although the most common locations are the extremities, it should be kept in mind that it can occur in any part of the body (3,4). Patients with extremely rare intrapelvic SS; may present with acute urinary retention, pain in the pelvic region, constipation, and lower urinary tract symptoms. In the presented case; benign prostatic hyperplasia was considered in the preliminary diagnosis of the patient who developed acute urinary retention and presented with lower urinary tract symptoms; SS was determined by imaging and histopathological examination of the mass removed after pelvic exploration.

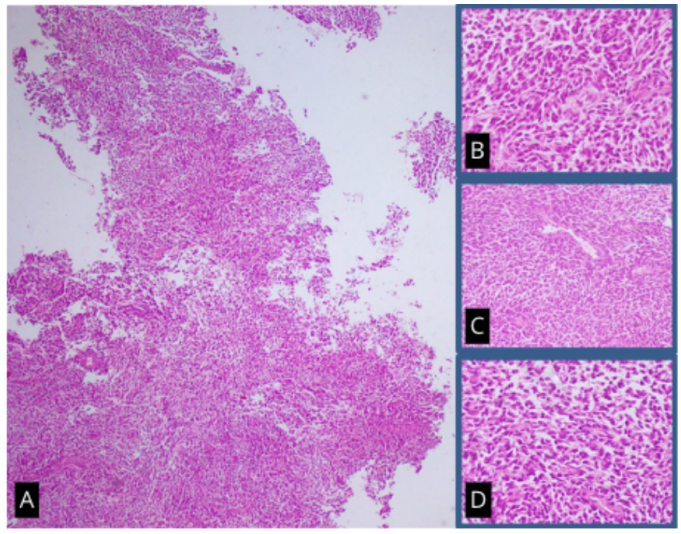

Figure 2. A. At small magnification; tumor cells with a spindle-like distribution, monotonous, hyperchromatic nuclei and narrow cytoplasm are observed (H&E X40). B. Among the tumor cells with spindle, hyperchromatic nuclei and narrow cytoplasm, an area where mitotic active cells can be selected is observed (H&E X400). C. Tumor cells with spindle-like patterns lined up around the vascular structure are seen (H&E X200). D. There is an area where the spindle structures of the tumor cells can be clearly observed (H&E X400)

Histologically, the tumor may be monophasic or biphasic, with varying proportions of epithelial and spindle cells. Specific immunohistochemical markers; may indicate vimentin and cytokeratin. In 90% of cases, there is a specific fixed translocation, usually a balanced reciprocal translocation in the form of t (X; 18) (p11.2;q11.2) (1,2). However, the presence of the said translocation was not investigated in the case examined. On CT examination, it typically presents as a soft tissue mass with slightly higher attenuation than muscle tissue, and there may be infiltration into adjacent tissues. While a heterogeneous density structure is observed, cystic density lesions containing fluid-fluid levels corresponding to bleeding areas can be rarely detected. The tumor usually shows a heterogeneous enhancement.

T1 and T2-weighted MRI usually show the heterogeneous signal intensity and may contain varying amounts of septation. Hyperintensity detected on T1 and T2-weighted images corresponds to bleeding areas. Fluid-liquid levels can be detected in 10-25% of cases (6,7,8). Mixed-signal appearance is detected in MRI examination in approximately one-third of the cases. In this case; in pelvic MRI examination; a giant mass lesion with a heterogeneous internal structure was observed, located in the midline in the pelvic region, with a lobulated contour, approximately 14x13 cm in size, pushing the bladder anteriorly and the rectum posteriorly, containing cystic and contrasting solid areas. At the level of the pelvic floor, it was observed that the mass pushed the prostate of normal size forward, resulting in the displacement of the prostate toward the symphysis pubis. It was noted that the mass was separate from the prostate but closely adjacent to the posterior prostate capsule. An extraprostatic pelvic mesenchymal tumor or tumor originating from the prostate capsule was considered in the radiological differential diagnosis.

Soft tissue sarcomas such as fibrosarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma, malignant schwannoma, which may contain calcification, and malignancies such as hemangiopericytoma and lymphoma, which are observed as lesions in soft tissue density or attenuation on CT and MRI, often in which low-density or attenuated areas of necrosis are observed; it shows radiological features similar to SS. Although the radiological findings described in this study can be detected in other malignant tumoral lesions, which are more common, in the presence of the defined imaging findings, SS must be considered in the differential diagnosis.

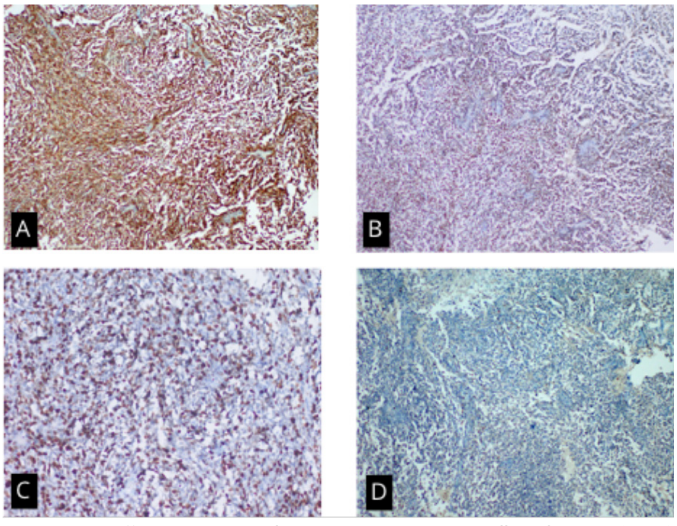

Figure 3. A. Diffuse strong cytoplasmic staining in tumor cells with BCL-2 (x100) B. Diffuse strong cytoplasmic staining in tumor cells with TLE (x100) C. 50% proliferative activity was observed in tumor cells with Ki-67 (X200) D. Diffuse pale cytoplasmic staining (x100) in tumor cells with EMA

EMA: Epithelial membrane antigen

The preferred treatment method in SSs; wide radical excision alone or along with radiotherapy. Healing is closely related to how radical the excision is. Tumor size, mitotic rate, and extensive tumor necrosis are considered as the most important prognostic determinants. Recurrence can be seen.

The incidence of SS was increasing day by day. We present a very rare case of intrapelvic SS causing acute urinary retention. This soft tissue tumor has a poor prognosis and may be confused with benign prostatic hyperplasia in the preliminary diagnosis, as it causes lower urinary tract symptoms (decreased urinary flow, incomplete emptying of the bladder, acute urinary retention, etc.). It is of great importance to report such cases for a better understanding of their pathophysiology and treatment options.

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Ethics

Informed Consent: Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: H.E.D., Design: Y.Y.K., Supervision: H.E.D., Data Collection or Processing: F.E.G.S., Analysis or Interpretation: F.D.A., Literature Review: A.N., Critical Review: Y.Y.K., Writing: A.N.

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