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J Case Rep Images Oncology 2024;10(2):20–25.
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Unprecedented presentation of pelvic synovial sarcoma: A compelling case report

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ABSTRACT

Synovial sarcoma (SS) is a rare tumor that typically arises in various body regions, with its manifestation in the abdominal-pelvic area being exceptionally uncommon. However, despite its rarity in this location, it often metastasizes to the thorax. Despite its slow growth, it can be misdiagnosed as benign in less than 10% of cases. Diagnosis necessitates identification of the SYT-SSX fusion transcript, alongside immunohistochemical and cytogenetic assessments. Treatment involves surgical resection followed by radiotherapy for localized tumors, while chemotherapy is necessary for metastatic cases. Despite treatment, synovial sarcomas have a high recurrence rate, with about half recurring within two years. We report the case of a 73-year-old patient from sub-Saharan Africa who presented to the emergency room with obstructive symptoms, revealing a mass near the anus compressing the rectum upon imaging, causing an obstructive syndrome. Despite initial tumor resection, histological analysis identified a neurofibromatous origin. Twenty months later, recurrence prompted a hospital visit, revealing thoracic metastases necessitating surgery and chemotherapy. Subsequent analysis confirmed perianal synovial sarcoma. This case underscores the rarity of pelvic-anal synovial sarcoma and emphasizes the importance of early detection and proper management. Synovial sarcoma should figure prominently on the list of differential diagnoses of high-grade pelvic masses due to the importance of adjuvant chemotherapy.

Keywords: Immunohistology, MRI, Pelvic synovial sarcoma, Surgery

How to cite this article

Lahlou L, Ihssan HH, Kaoutar M, Hatim E, Assad EB, Soukaina A, Hounaida J, Youssef O, Rachida L. Unprecedented presentation of pelvic synovial sarcoma: A compelling case report. J Case Rep Images Oncology 2024;10(2):20–25.

Article ID: 100137Z10CL2024

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doi: 10.5348/100137Z10CL2024CR

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Received: 15 March 2024
Accepted: 09 May 2024
Published: 16 October 2024

INTRODUCTION

Synovial sarcoma is characterized as a high-grade malignant tumor. Despite its potential to occur at any age ranging from 5 to 87 years, it is most frequently observed between the ages of 15 and 35 [1, 2]. Contrary to its name, these lesions typically develop near joints rather than within them. Predominantly, they manifest in the extremities, with reports of abdominal and pelvic occurrences comprising only 3–7% of cases [2, 3]. Synovial sarcoma is categorized as an intermediate to high-grade tumor with a significant propensity for metastasis. Given its aggressive nature, thorough pathological and

radiological evaluations are imperative for staging and determining the extent of the lesion to guide appropriate therapy. While imaging features are not pathognomonic, they often provide valuable clues for diagnosis.

In cases where margins are involved or closely positioned, a recommended therapeutic approach involves comprehensive local excision of the tumor, often complemented by radiotherapy. While adjuvant chemotherapy is advisable in cases with secondary localization [4].

Within this framework, we present the case of a 73-year-old male patient whose diagnosis was confirmed through immunohistochemical staining. The objective of this case report is to enhance understanding, diagnosis, and treatment of perianal synovial sarcoma.

CASE REPORT

We present the case of a 73-year-old man who had well-controlled type I diabetes and had been effectively treated for pulmonary tuberculosis since 2019, and was certified cured. His illness began one year ago when he presented with dysuria, bowel transit disturbances complicated by acute intestinal obstruction, resulting in weight loss, and abdominal distension, prompting a visit to the emergency department in his home country, where he was admitted to the operating room.

Eight months later, he presented to our National Oncology Institute with a recurrence of previously mentioned digestive and urinary symptoms, but no surgical report was provided. Only a letter stated that the patient's tumor had been completely removed, and the histological study revealed that the tumor was caused by neurofibromatosis.

Subsequently, we conducted a contrast-enhanced thoraco-abdominopelvic computed tomography (CT) scan revealing a pelvic tissue mass measuring 80 × 120 × 160 mm with heterogeneous density containing a lateralized necrotic area on the right side. It infiltrates the anal margin from behind, involves the pelvic peritoneal layers on the left, and exerts a mass effect on the rectum. Anteriorly, it comes into close contact with the posterior wall of the bladder. Laterally, it infiltrates the internal obturator muscles and approaches the iliac vessels. Additionally, there was a 10 mm pre-sacral lymph node and 8 mm internal iliac lymph node. In the thoracic region, a left peribronchovascular consolidation was identified, associated with suspicious-looking branched nodules in the lower left lobe (Figure 1).

A later magnetic resonance imaging (MRI) was performed showing a retroperitoneal and latero-rectal mass with heterogeneous signal on T2, containing some areas of necrosis enhanced heterogeneously after gadolinium injection with localized restriction on the diffusion sequence. Anteriorly, it exerts a mass effect on the bladder without affecting the upper urinary tract, and laterally, it comes into contact with the obturator muscles and the rectal wall without clear signs of infiltration, with the identification of a fistulous tract between the tumor and the soft tissues of the buttocks (Figure 2).

The patient underwent a new surgical procedure, and a resection of the perianal mass was performed, along with a colostomy, while preserving the anal canal. The operative specimen was transported to the pathology department for histopathological and immunohistochemical examination. Macroscopically, it appeared as an ill-defined and non-encapsulated mass weighing 2740 grams, measuring 20 × 19 × 10 mm. On sectioning, there was a whitish-gray indurated multilocular lesion occupying almost the entire specimen, with the presence of a muscle flap measuring 6 × 3 cm that appeared to be infiltrated.

Microscopically, the histological examination of the analyzed sections reveals a tumoral proliferation composed of spindle-shaped cells. These tumor cells have elongated, anisocaryotic, and hyperchromatic nuclei with nucleoli. The cytoplasm appears eosinophilic. The mitotic index is 5 mitoses per 10 high-power fields. There is the presence of tumor necrosis estimated to be more than 50%, and no vascular embolism is associated. The tumor infiltrates the surrounding adipose and striated muscle tissue at the periphery, with the resection margin passing through the tumor itself (Figure 3).

The immunohistochemical markers were positive for anti-EMA, anti-AE1/AE3, and BCL-2 in more than 50% of the tumor cells, very weakly positive for anti-Ini1 (Figure 4) and negative for anti-PS 100, anti-smooth muscle actin, anti CD 34, anti-desmin, and anti-CD3 which is consistent with a diagnosis of synovial sarcoma (Figure 4).

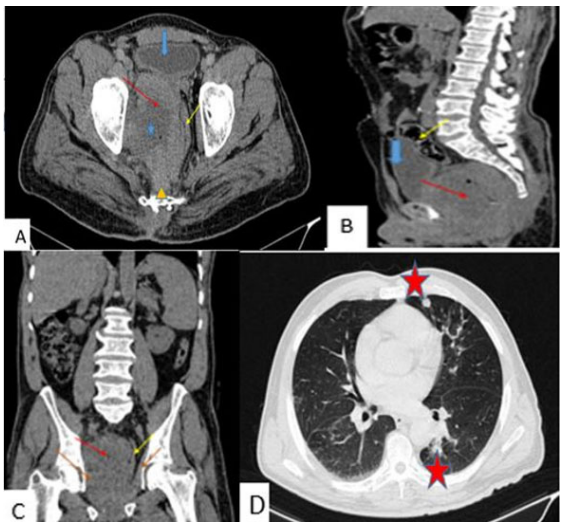

Figure 1: Abdominal CT: axial C+ section (A) sagittal and coronal section C–, (B, C) pelvic retroperitoneal multilobulated mass (red arrow) with necrotic area (blue star) exerting a mass effect on the rectum in the left (yellow arrow), on the anal canal below, on the bladder anteriorly (blue arrow), and on the obturator internus muscle laterally (orange arrow) with persistent fatty separation axial section in the pulmonary parenchymal window. (D) Metastatic pulmonary nodules in the lower left lobe (red star).

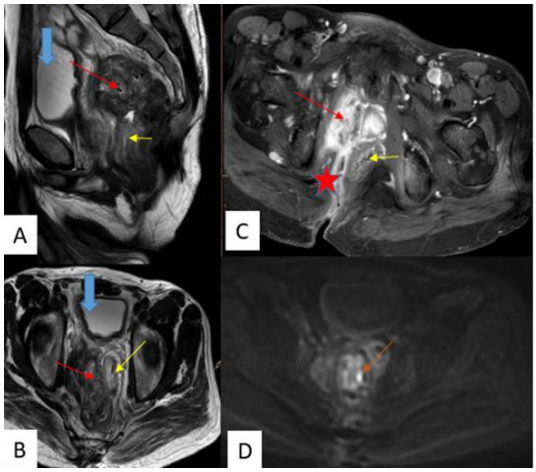

Figure 2: Abdominal MRI: Sagittal and axial T2 (A, B): retroperitoneal latero-recto-anal multilobulated mass with heterogeneous T2 hypointensity (red arrow), creating a mass effect on the left rectum (yellow arrow) and anal canal, and on the bladder anteriorly (blue arrow), coming into contact with the internal obturator muscle without clear signs of infiltration. T1 after injection of gadolinium (C), and diffusion B1000 (D). This process enhances heterogeneously after gadolinium injection (red arrow), with the identification of a fistulous tract between the tumor and the soft tissues of the buttocks (red star). Localized restriction is also observed on the diffusion sequence (orange arrow).

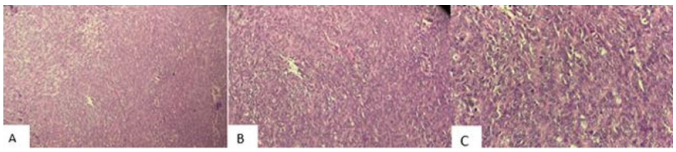

Figure 3: Low (A)/medium (B), and high (C) magnification: histological appearance of a spindle cell tumor.

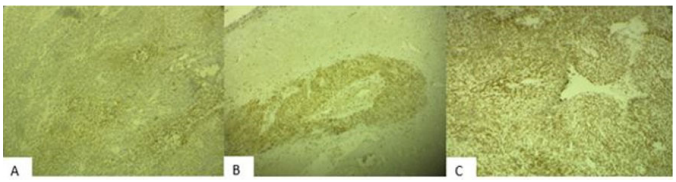

Figure 4: Immunohistochemical study: positive expression of AE1, AE3, and EMA antibodies in more than 50% of the tumor cells (A, B). Weakly positive expression of Ini1 antibodies (C).

The interpretation of the fluorescence in situ hybridization results using a break-apart probe showed SYT translocation. EWSR1 translocation was absent.

In conclusion, the morphological and immunohistochemical features were suggestive of monophasic synovial sarcoma associated with poorly differentiated areas. The patient was subsequently referred to our oncology department for continued follow-up. He received a course of ifosfamide-doxorubicin treatment.

DISCUSSION

Synovial sarcoma (SS) is an uncommon tumor constituting approximately 7–8% of all mesenchymal malignancies, with diagnoses typically occurring between the ages of 26 and 38, though cases occur across a wide age range. While slightly more prevalent in males, SS predominantly manifests in the extremities, with primary thoracic occurrences being rare and diverse, affecting various organs such as the heart, lungs, stomach, esophagus, mediastinum, and thoracic wall in about 8% of cases. Within this spectrum, primary intra-abdominal synovial sarcoma is exceptionally rare, often presenting in the retroperitoneal region, albeit sometimes disguised due to its size.

Though officially accounting for about 1% of retroperitoneal malignancies, the true prevalence of synovial sarcoma in this region may be underestimated, as some cases previously classified differently might be reclassified upon closer examination. Intra-abdominal and pelvic SS typically arise in locations like the mesentery, gastrocolic ligament, and inner aspect of the abdominal wall, with occasional involvement of the omentum.

Pelvic tumors, except those originating from skeletal or related limb girdle components, are uncommon, with some pelvic tumors being classified as retroperitoneal due to their proximity to the pelvic diaphragm's levator muscles.

Clinically, patients with pelvic synovial sarcoma present with symptoms such as pelvic heaviness, vague digestive discomfort, and pain during defecation, due to the tumor's pressure on the canal anal and the rectum [5].

In terms of appearance, synovial sarcoma can be divided into three basic histologic subtypes: biphasic, monophasic, and poorly differentiated [6].

When studied under light microscopy, biphasic synovial sarcoma consists of a mesenchymal spindle cell and a separate epithelial component.

Monophasic synovial sarcoma is the most common subtype, accounting for 50–60% of intra-abdominal and pelvic cases. In this subtype, mesenchymal spindle cells take priority. These spindle-shaped cells are unattractive, with ovoid, pale-staining nuclei and little visible nucleoli. They grow in a manner similar to interlaced bundles and exhibit low to moderate cell division activity. Notably, the surrounding tissue, or stroma, is often pinkish, separating it from malignant peripheral nerve sheath tumors, which appear deeper blue (at lower magnification). It is worth emphasizing that finding monophasic epithelial tumors may be challenging without genetic evidence.

Poorly differentiated synovial sarcoma has more highly pigmented nuclei that mimic microscopic round cell sarcomas or carcinomas. Except for plumper cells clustered in some tumors, there is little variation in cell shape and cytoplasm. Significant myxoid changes in tissue are uncommon [4–6].

While synovial sarcoma has a variable immunohistochemical profile, the majority of these

tumors test positive for cytokeratin (CK7 and CK19) and epithelial membrane antigen (EMA), which are virtually 100% specific. Recent research has shown that EMA, cytokeratin AE1/AE3, and E-cadherin, when combined with the absence of CD34 expression, are the most reliable and sensitive markers for diagnosing monophasic fibrous and poorly differentiated synovial sarcoma.

Approximately 80% of synovial sarcomas include the reciprocal chromosomal translocation t(x;18) (p11.2;q11.2). Monophasic tumors often feature rearrangements involving either SSX1 or SSX2, whereas the majority of biphasic tumors have the SSX1 rearrangement. In a recent study, only 3% of biphasic tumors tested positive for SYT-SSX2 [7].

The radiographic characteristics of synovial sarcomas may not be entirely clear, but certain patterns are commonly observed on imaging. Typically, on CT scans, synovial sarcomas appear as heterogeneous, deep-seated tumors with attenuation levels similar to muscle. Areas of lower attenuation, indicating necrosis or bleeding, are often present. These tumors often exhibit a multinodular morphology with either well-defined or irregular borders. Following intravenous contrast material administration, synovial sarcomas typically show heterogeneous enhancement, aiding in their differentiation from cystic lesions or hematomas seen on pre-contrast imaging [8].

Computed tomography (CT) scans are particularly useful for detecting calcifications, present in a significant portion of synovial sarcomas, and for determining bone involvement, especially in complex anatomical locations like the pelvis.

However, for staging and diagnosis, MRI is the preferred radiological approach, providing a more detailed assessment of the disease's extent and intrinsic characteristics. On T1-weighted MR scans, synovial sarcomas often appear as heterogeneous, multilobulated masses with signal intensity comparable to or slightly greater than muscle. T2-weighted MRI typically shows significant heterogeneity and a "triple sign," indicating a combination of solid cellular elements, areas of hemorrhage or necrosis, and regions with calcification or fibrotic collagenization.

Post-contrast injection, synovial sarcomas often exhibit considerable enhancement, with diffusion patterns varying depending on tumor viability. Magnetic resonance imaging findings suggesting high-grade synovial sarcoma include the absence of calcification, the presence of cystic components, evidence of bleeding, and recognition of the "triple sign" [9].

Following chemotherapy or radiation therapy, T2-weighted MR scans may show increased signal intensity within the tumor, indicating increasing necrosis, and may also reveal a reduction in tumor size. However, post-treatment modifications may not always be visible on images with short echo durations [10].

Differentiating intra-abdominal and pelvic synovial sarcoma from other illnesses frequently necessitates considering a variety of possible diagnoses, such as biphasic neoplasms, spindle cell sarcomas, and small round cell tumors.

Biphasic synovial sarcomas must be distinguished from carcinosarcoma, Wilms' tumor, extragonadal germ cell tumors, and mesotheliomas. Carcinosarcomas often develop in gynecological organs in older people, with greater pleomorphism and cytokeratin positive than synovial sarcomas.

Similarly, monophasic synovial sarcoma can resemble a variety of different neoplasms, including solitary fibrous tumors, malignant peripheral nerve sheath tumors, leiomyosarcomas, and gastrointestinal stromal tumors arising in the retroperitoneum or omentum. The differentiation is mostly based on differential physical traits and immunophenotype.

For poorly differentiated synovial sarcomas, their similarity to other small round cell sarcomas may demand the use of immunohistochemistry panels and genetic data for proper differentiation [11].

Surgical excision remains the primary treatment for synovial sarcoma, regardless of its location in the body, in cases without metastasis. Patients with metastatic disease are typically treated with ifosfamide and/or doxorubicin as first-line therapy. However, the prognosis may vary depending on factors such as tumor size, histological grade, and SSX2 gene rearrangements. Vigilant monitoring is essential due to the likelihood of recurrence, particularly when resection margins are unclear [12, 13].

CONCLUSION

In conclusion, our patient underwent a complete tumor resection followed by adjuvant chemotherapy given the secondary lung localization. Since the resection margins were not clear and extended into the tumor itself, the likelihood of recurrence is inevitable, underscoring the importance of vigilant monitoring.

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Author Contributions

Chaimae Lahlou – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Hadj Hssain Ihssan – Conception of the work, Design of the work, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Meslouhi Kaoutar – Conception of the work, Design of the work, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Essaber Hatim – Conception of the work, Design of the work, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

El Bakkari Assad – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Allioui Soukaina – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Jerguigue Hounaida – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Omor Youssef – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Latib Rachida – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

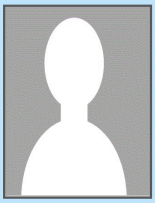
Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

ABOUT THE AUTHORS

Article citation: Lahlou L, Ihssan HH, Kaoutar M, Hatim E, Assad EB, Soukaina A, Hounaida J, Youssef O, Rachida L. Unprecedented presentation of pelvic synovial sarcoma: A compelling case report. J Case Rep Images Oncology 2024;10(2):20–25.



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