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Case Report

Triple Local Recurrence of Synovial Sarcoma: A Case Report

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ABSTRACT

Background: Synovial Sarcoma (SS) hardly appears on non-extremity sites (20% of cases). Even rarer is a local recurrence, which occurs between 10-30% of cases. Here, we present a rare case of hip Synovial Sarcoma, which recurs three times.
Case Report: A 33-year-old male came to our outpatient clinic with chief complaints of a sudden ingrowing mass on his left hip. The lump is the size of a chicken egg and doesn't get bigger over time. There was palpable tenderness felt whenever he touched the lump. The patient was diagnosed with SS of the left hip with IIB stage from additional radiographic and histopathologic examination. Surgical-wide resection had been done three years before the visit, and chemotherapy six times. The lump reappeared one year before the visit, and the patient underwent a second surgery. The mass re-surfaced within the same region on the current visit for the third time, and he underwent the same surgical treatment afterward. Two years after the third surgery and adjuvant chemotherapy, the patient had no complaint about a lump and planned a radiograph evaluation.
Discussion: Detailed examination from history taking, physical examination, histopathologic, and radiological results is important to determine appropriate staging. With proper staging, physicians can better understand disease prognosis and risk of recurrence or metastases.
Conclusions: The third recurrence may still happen on SS. Its unique location might contribute to this unique occurrence, and the size of the tumor makes the recurrence rate higher.

Keywords: Synovial sarcoma; Malignancy; Recurrence; Human and medicine

INTRODUCTION

Synovial sarcoma is a very uncommon malignant neoplasm that manifests in roughly 6-9% of adults diagnosed with soft tissue sarcomas. Synovial sarcoma comprises approximately 5 - 10% of all soft tissue sarcomas.[1,2] Synomial Sarcoma has the potential to manifest in several anatomical locations. However, a significant proportion of cases are observed in the extremities.[3] While synovial sarcomas have the potential to form in periarticular areas, they rarely originate within the joint and are not connected to typical synovial tissue.[4] The trunk, retroperitoneal/abdominal, and head and neck regions are the most often observed non-extremity sites, accounting for 8%, 7%, and 5%, respectively. The phenomenon of SS has the potential to exhibit recurrence. The occurrence of local recurrence exhibited variability across the literature, with reported rates ranging from 10% to 30%.[5] The recommended approach for managing SS is the total excision of the tumor with a negative resection margin, along with peri-operative radiation therapy if deemed necessary. Nevertheless, the efficacy of adjuvant chemotherapy remains a subject of debate. It has solely been validated through retrospective analysis to enhance survival rates in patients with a high-risk profile.[6] Several important prognostic markers have been identified in the literature. These factors include tumor size greater than 5 centimeters, deep-seated tumor localization, the adequacy of surgical margins, and a history of recurrence.[7]

Given its rare occurrence in non-extremity sites and unlikeliness in recurrence, here we presented a rare case of SS where the SS occurs on the hip and had local recurrence three times.

CASE REPORT

A 33-year-old male came to our hospital outpatient clinic with chief complaints of a sudden ingrowing mass on his left hip. There was no history of any traumatic injury near the lump. The lump is the size of a chicken egg and doesn't get bigger over time. There was palpable tenderness felt whenever he touched the lump. From the history taking, information about how he used to come to the outpatient clinic of a surgeon in Lumajang General Hospital and then referred to our hospital for further diagnostic and therapeutic evaluation is obtained. By November 2016, the patient had undergone surgical-wide resection in our hospital. From January until July 2017, he received chemotherapy treatment six times. After the first surgery, the patient has no complaint about relapse mass. By 2019, the egg-sized mass re-surfaced again in the same region, and the wide excision was done by October 2019. The mass re-surfaced within the same region for the third time in 2021, and he underwent the same surgical treatment by November 2021. Two years after the third surgery and adjuvant chemotherapy, the patient has no complaint about a lump and is planned for radiograph evaluation. The same orthopaedic oncologist did all the surgery. The consent for the case reporting has already been given to the patient.

From the physical examination, we found a scar on the left hip with a lump of mass nearby in the left inguinal region the size of +/- 7cm. The lump has no color change, venectation, or other appearance changes. This mass felt painful when touched and moved. There is no problem with the distal neurovascular evaluation. Both active and passive hip range of movement is not limited.

Radiological evaluations were done to understand the problem. The first MRI of the pelvis at the 2016 pelvic was suggestive of malignant intramuscular mass towards the hip flexor muscle group, with an average diameter of 12 cm and no infiltration to subcutaneous tissue and vascular encasement of a/v femoral (Figure 1). After the first surgery and six times chemotherapy, the Left femur MRI evalu-

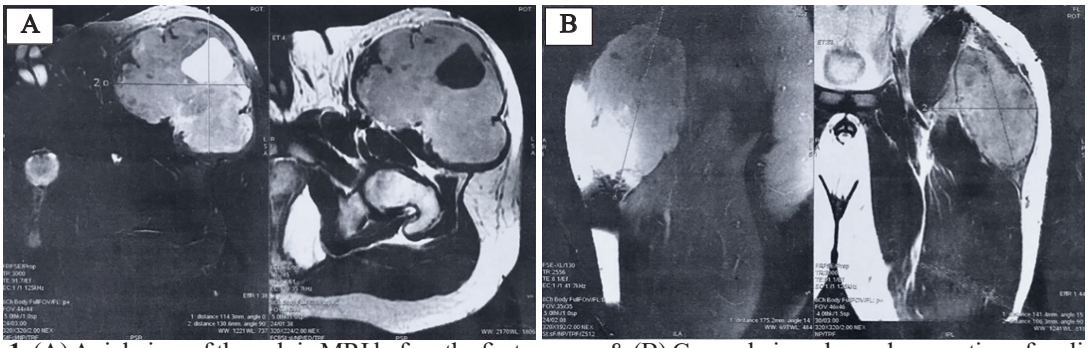

Figure 1. (A) Axial view of the pelvic MRI before the first surgery & (B) Coronal view showed suggestive of malignant intramuscular mass

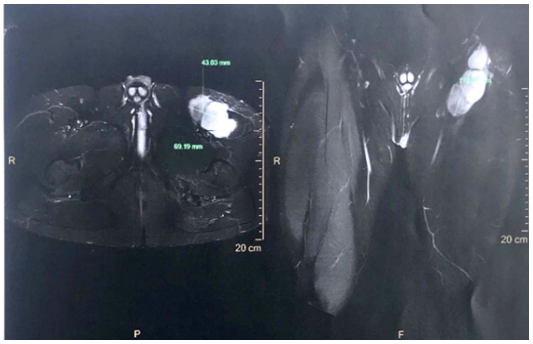

Figure 2. (A) Axial view of the femur MRI evaluation two years after first surgery & (B) Coronal view showed suggestive recidive sarcoma

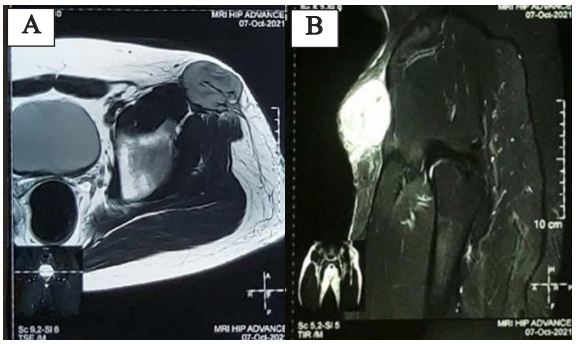

Figure 3. (A) Axial view of hip MRI before the third surgery & (B) Coronal view showed recidive mass at the surgical bed of the left proximal femur.

ation in 2018 showed no recidive mass at the surgical bed. At the follow-up, the Lower extremity MRI 2019 with contrast showed a recidive mass at the surgical bed that infiltrated the rectus femoris and lateral vastus muscles (Figure 2). The second surgery was performed in 2019, followed by chemotherapy and radiotherapy. The evaluation in 2021 showed a suggestion of a malignant Intramuscular mass infiltrating the proximal femur region (Figure 3) and chest x-ray AP and lateral showed no lung metastases (Figure 4).

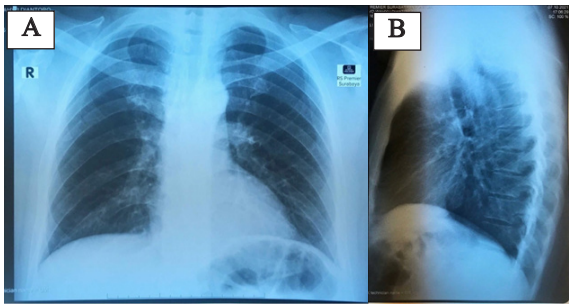

Figure 4. (A) Chest x-ray AP and (B) Lateral showed no lung metastases.

Pathological examination of the tumor tissue obtained from the first surgery showed a mass with size 11 x 13x 14 cm, and histopathologic examination indicated a malignant spindle mesenchymal tumor with the differential diagnosis of grade pleiomorphic undifferentiated sarcoma (MFH), fibrosarcoma, and Synovial sarcoma with a margin of the excision was free from tumor (Figure 5). IHC was done with EMA, CK. Vimentin was positive, which suggests synovial sarcoma (Figure 6). Several histopathologic examinations following the next surgical treatment showed synovial sarcoma as well. This case is then assessed as recidive synovial sarcoma of the left hip with IIB stage.

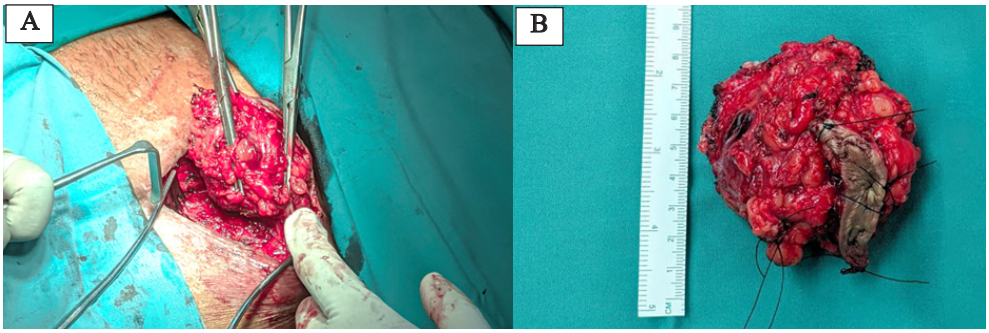

Figure 5. (A) wide excision is done to remove the mass of the left hip and (B) the tumor tissue obtained from the first surgery showed a mass with size 11 x 13x 14 cm.

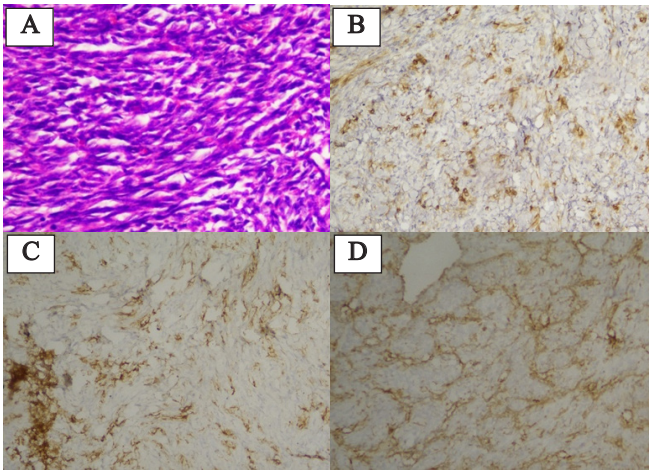

Figure 6. (A) Staining of H&E, (B) IHC of EMA positive, (C) CK positive, (D) Vimentin suggest synovial sarcoma

DISCUSSION

Patients with synovial Sarcoma exhibit clinical behavior and outcome variations. Synovial sarcoma is widely acknowledged as a high-grade sarcoma characterized by severe prognosis. The anticipated 5-year survival rate in adult patients ranges from 50% to 60%, while the 5-year metastatic disease-free survival (DFS) ranges from 40% to 60%. Nevertheless, there are documented cases exhibiting a passive demeanor, wherein patients may encounter a recurrence of symptoms even a decade after first diagnosis.

The diagnosis of synovial sarcoma can be achieved with a comprehensive approach involving the assessment of medical history, physical examination, magnetic resonance imaging, biopsy, subsequent pathological examination, immunohistochemistry, and molecular analysis. Similar to other surgical procedures in the field of Surgical and Technical Sciences (STS), it is recommended that a biopsy be conducted before decisive surgery to prevent incomplete removal of tissue and incorrect diagnosis. Various biopsy options comprise open biopsies, core needle biopsies, and fine needle aspirations (FNA). Compared to alternative methods, incisional biopsy demonstrates superior diagnostic accuracy; however, it is associated with a higher incidence of complications than the closed procedure.[9,10]

Staging investigations are significant as they provide a more comprehensive illness prognosis and chances of recurrence or metastasis. The tumor stage is also crucial for treatment strategy by a multidisciplinary team specializing in sarcoma. The staging for synovial sarcoma includes the utilization of cross-sectional imaging of the affected extremity, a systematic staging involving a chest CT scan, and a pathologic examination. Synovial sarcomas exhibit malignancy and have the propensity to metastasis, primarily to the lungs. It has been shown that approximately 13% of patients report distant metastases at the time of diagnosis.[11,12]

The European Pediatric Soft Tissue Sarcoma Study Group recently presented results indicating a more favorable prognosis in a subset of pediatric patients with synovial sarcoma, as seen by a 90% 5-year survival rate.[8] The size of a tumor has been identified as an indicator of survival in most studies. Moreover, the prognosis can be influenced by the tumor's location, as cancers originate from anatomical areas other than the extremities. Histology-specific nomograms for synovial sarcoma are accessible, which include factors such as patient age, tumor site, size, and grade. The current standard procedure for managing primary, localized synovial sarcoma entails the extensive surgical removal of the tumor in addition to radiation therapy. This treatment strategy is particularly recommended for high-risk patients, including grade 3 tumors, deep tumor location, and tumor size of 5 cm or greater. Cytotoxic treatment is frequently considered for patients with advanced SS in neoadjuvant and adjuvant settings. Furthermore, ongoing research is being conducted to explore novel molecular agents and immunologic approaches.

The recurrence tumor of this case can be caused by a large tumor of more than 5 cm from the first surgery, although pathologic evaluation suggests a free margin of the tumor. After years of evaluation, synovial sarcoma can still be reemerged at the same site. The strategy to manage the recurrency consists of a multidisciplinary section with a surgeon, medical oncologist, radiologist, pathologist, and nuclear medicine. The treatment includes adjuvant radiation and chemotherapy.

CONCLUSION

The third recurrence may still happen on SS. Its unique location in the pelvic region might contribute to this unique occurrence, and the tumor size makes the recurrence rate higher.

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None

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