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Case Report

A rare case of perineal synovial sarcoma ☆,☆☆

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ABSTRACT

Synovial sarcoma is a malignant mesenchymal neoplasm that is frequently misdiagnosed due to its slow growth and small size. This tumor presents as a nonspecific heterogeneous mass on cross-sectional imaging. Biopsy and histopathological assessments are required to differentiate synovial sarcoma from other sarcoma subtypes and to define the tumor grade. This article presents the case of a 17-year-old male patient with perineal synovial sarcoma.

Introduction

Soft tissue sarcomas (STS) are a special subgroup of sarcomas, and synovial sarcomas make up 5%-10% of all STS [1]. The typical age at presentation is 32 years, and both men and women are equally affected. The most frequent location for synovial sarcoma is the extremities, and 70% of cases involve the lower extremities. Synovial sarcomas typically develop in the popliteal fossa; however, this malignancy can also affect the hand, wrist, upper extremity, and proximal limb girdle [2]. To our knowledge, there are relatively few reports in the literature of synovial cancer in the buttocks. In this article, we present the case of a 17-year-old male patient whose condition was identified via immunohistochemical staining. The purpose of this report is to improve the understanding, diagnosis, and management of synovial sarcoma.

Case description

A 17-year-old male who had no relevant medical history or family health history presented at our hospital due to

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the growing mass in his right buttock over approximately 6 months. He had local excision at a nearby hospital in December 2020 when it was discovered that he had a somewhat uncomfortable subcutaneous tumor in the right buttock. At the local hospital, the patient was first identified as having a benign tumor of the ischium, and the tumor was surgically removed. As a result, the patient had simply surgical excision of the tumor and no other treatments. Physical examination revealed nothing more than a lump in the right buttock that was around 5 cm in size and showed no symptoms of inflammation.

His blood test results were in the normal range. Magnetic resonance imaging (MRI) revealed 2 masses located in the subcutaneous fat layer on the right side of the perineum. The masses consisted of a peripheral area that was strongly enhanced after injection and a fluid part in the center that did not enhance. The markedly heterogeneous appearance of these tumors on fluid-sensitive sequences includes areas of necrotic and cystic degeneration with very high signal, soft tissue components with a relatively high signal, and areas of low signal intensity due to dystrophic calcifications and fibrotic bands. The MRI scan did not reveal any relationship between the masses and the anus or rectum, and no invasion of adjacent bones, muscles, or skin was noted (Fig. 1). An initial diagnosis of soft tissue neoplasm was made.

The patient underwent local excision at our hospital. Macroscopically, the tumor was identified in the right buttock. Two solid tumors with dimensions 3 × 4 cm and 2 × 3 cm, joined together like a dumbbell (Fig. 2), were well demarcated with surrounding tissue and were located below the levator anal muscle from the right lateral rectus fossa to the base of the scrotum.

Microscopically, these tumors contained monotonous spindled cells (Fig. 3A) and a hemangiopericytoma-like vascular pattern (Fig. 3B). They exhibited diffuse TLE1 (Fig. 3C), CD99 (Fig. 3D), vimentin (Fig. 3E), and BCL2 (Fig. 3F) expression. Based on these results, monophasic synovial sarcoma was diagnosed. The patient was subsequently referred to our cancer center for continued follow-up. He received anthracycline-based chemotherapy with the addition of ifosfamide. The patient died within 1 year after surgery.

Discussion

Soft-tissue sarcoma is a rare cancer that accounts for approximately 1% of all malignant tumors. Although they occur in various age groups, soft-tissue sarcomas account for 8% of all malignant tumors developing in adolescents and young adults, suggesting that they are not rare in this age group [3].

Synovial sarcoma is a type of soft-tissue sarcoma, comprising 5%-10% of all STS [1]. According to the Bone and Soft-Tissue Tumor Registry in Japan, synovial sarcomas occur predominantly in adolescents and young adults among STS [4]. Whereas, the major histologic subtypes of STS including undifferentiated pleomorphic sarcoma (UPS), well-differentiated liposarcoma (WDLS), leiomyosarcoma (LMS), myxofibrosarcoma (MFS), malignant peripheral nerve sheath tumor (MPNST), and dedifferentiated liposarcoma (DDLS) had a peak in

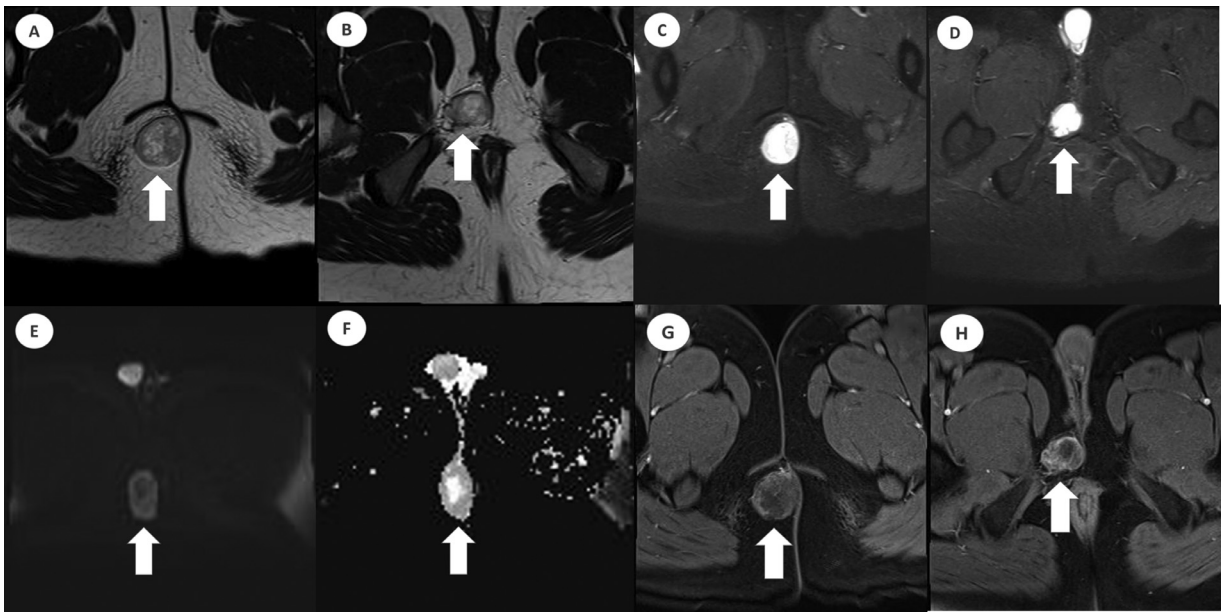

Fig. 1 – Two masses (arrows) with well-defined borders located on the right of the perineum had mixed-intensity signals on T2-weighted imaging (A, B) with the presence of solid cellular elements (intermediate signal intensity), necrosis (high signal intensity), and fibrotic regions (low signal intensity); the tumors (arrows) had high intensity on T2-weighted fat-saturated images (C, D); the solid peripheral area exhibited significant restriction on diffusion-weighted imaging, and the fluid central part had unrestricted diffusion (arrows) (E, F); on dynamic contrast-enhanced T1-weighted fat-saturated images, the tumors(arrows) showed enhancement (G, H).

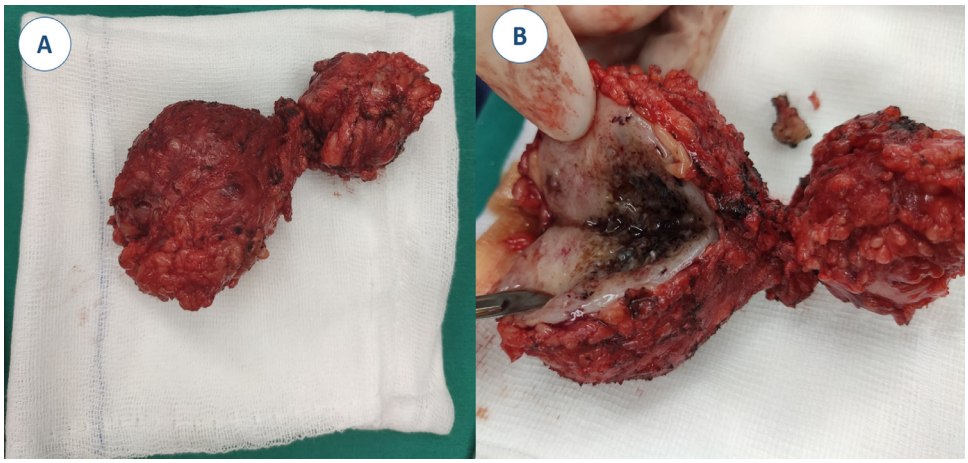

Fig. 2 – Two solid tumors with dimensions 3 x 4 cm and 2 x 3 cm joined together like a dumbbell.

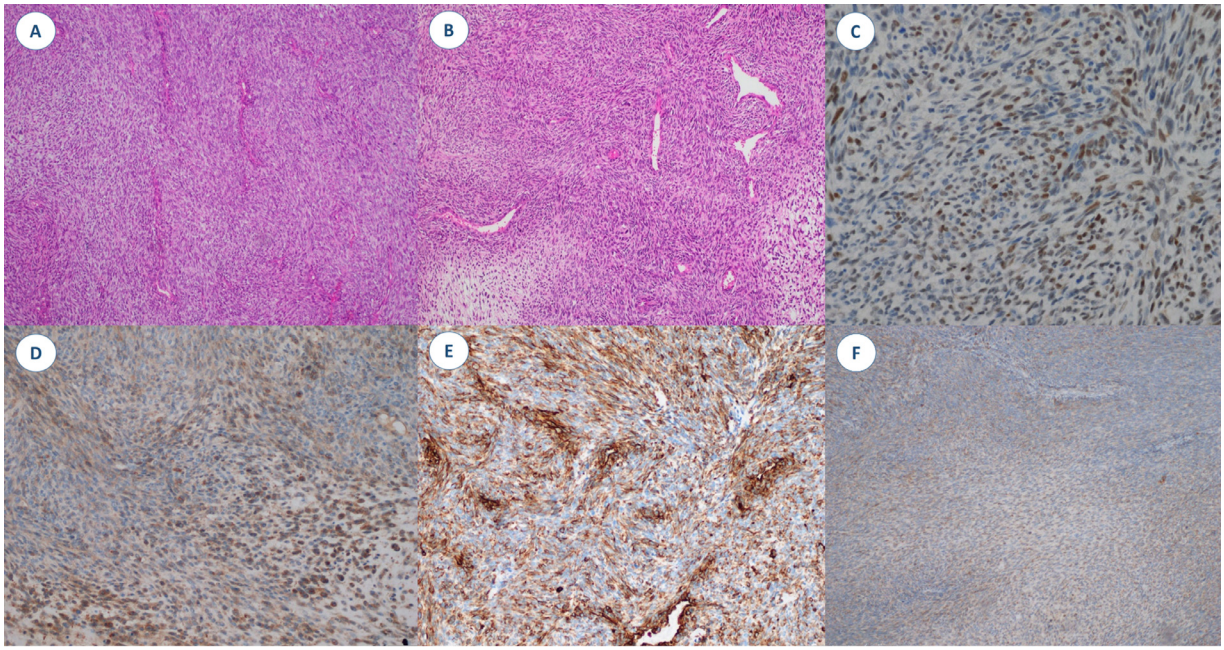

Fig. 3 – Histological findings of the tumors in the described patient with monophasic synovial sarcoma. Image A and B illustrate hematoxylin and eosin staining, x 10 and x 20, respectively. Immunohistochemistry was positive for TLE1 (C), CD99 (D), vimentin (E), and BCL2 (F).

the elderly [4]. The term "synovial sarcoma'' is a misnomer because these tumors do not originate from the intra-articular synovium but rather from primitive mesenchymal cells; however, many of these tumors develop close to articular structures [5]. A chromosomal aberration called T (x; 18) (P11.2; q11.2), which results in the production of the SS18-SSX fusion oncogene, is a broadly accepted etiology of synovial sarcoma. It was detected in more than 90% of cases imparting definitional significance [6].

Synovial sarcomas often do not present with the typical STS presentation of a large and quickly growing painless mass [7]. As an alternative, the majority of synovial sarcomas develop slowly. These individuals may initially receive a clinically incorrect diagnosis due to the gradual onset, younger age at presentation, and unusual presenting symptoms. Due to its good tissue contrast and ability to visualize the lesion in many planes, MRI is the recommended imaging modality for the assessment of STSs. It can be used to assess the tumor's size and the extent to which it has affected nearby soft tissue structures, distinguish the tumor from muscle tissue and demonstrate the involvement of neurovascular systems, tendons, fascial/fat planes, and bone marrow. Syn-

ovial sarcoma lesion margins are clearly defined on MRI in 53%-91% of cases and poorly defined in 9%-47% of cases. Small size, well-defined borders, and uniform appearance on cross-sectional imaging are the characteristics of synovial sarcoma that may result in an initially incorrect diagnosis of benign indolent disease. Although these tumors' radiographic characteristics are not pathognomonic, the presence of a soft-tissue mass in a young patient that is close to but not in a joint, especially if it is calcified (30%), is highly indicative of the diagnosis [8]. On T2-weighted MRI, the predominant high signal intensity with apparent heterogeneity is also a key characteristic of synovial sarcoma. The "triple sign'' (low, intermediate, and high signal intensity areas) is a term used to describe this nonhomogeneity in the signal. It denotes the simultaneous presence of solid cellular components (intermediate signal intensity), hemorrhage or necrosis (high signal intensity), and fibrotic regions (low signal intensity). In between 35% and 57% of synovial sarcoma cases, and more frequently in bigger tumors, the "triple sign'' can be found. Many other soft-tissue neoplasms also have the "triple sign'' as an imaging characteristic. Among them is fibrous histiocytoma [9]. In patients affected by synovial sarcoma, the triple sign that is identifiable on MRI (T2-weighted sequences) is associated with decreased disease-free survival. Furthermore, individuals with synovial sarcoma without calcifications have poor disease-free survival [10]. Roughly 30% of patients have radiologic calcifications that can be seen. These calcifications can be focal or widespread throughout the tumor, and they frequently have a fine, stippled, or opaque appearance [11].

In order to distinguish synovial sarcoma from other STS subtypes and determine the tumor grade, a sample, and pathologic evaluation are necessary. Similar to other STS, a biopsy should be done before a final surgery to prevent an insufficient resection and a false diagnosis [12]. Incisional biopsies, core needle biopsies, and fine needle aspirations (FNA) are all available as biopsy options. The 3 main histological subtypes of synovial sarcoma are biphasic, monophasic, and poorly differentiated [13]. The biphasic subtype is characterized by the coexistence of epithelioid cells and spindle cells, whereas the monophasic subtype is composed solely of spindle cells [8]. BCL-2 expression is often widespread in immunohistochemistry, and CD99 staining is positive in 60% of cases. Furthermore, the transcriptional corepressor TLE1, which may be identified in most synovial sarcomas, exhibits significant and widespread nuclear staining on immunohistochemistry [14].

There are no established guidelines for managing synovial sarcoma. The ideal mode of treatment is broad surgical resection with tumor-free margins, followed by chemotherapy and/or radiation [15]. For cancers that have metastasized or are locally invasive, surgery is not advised. In rare circumstances, chemotherapy administered before and after surgery may be necessary [16,17]. The best course of treatment for each patient is decided in a multidisciplinary setting that considers both patient and tumor factors. A total of 150 individuals with nonmetastatic synovial sarcoma underwent radiation treatment and conservation surgery between 1960 and 2003. The median follow-up was 13.2 years; at 5, 10, and 15 years, the overall survival rates were 76%, 57%, and 51%, respectively [18]. The median survival time and 5- and 10-year survival rates were examined for each subtype of synovial sarcoma. The biphasic subtype had the most favorable prognosis, with a median survival of 30.75 years and the highest survival rates of 69% at 5 years and 60% at 10 years. In contrast, the epithelioid cell subtype had the lowest recorded 5-year survival rate (32%; 10-year survival rate: 6%; median survival: 1.25 years). Another report found that the median survival time and 5- and 10-year survival rates for the monophasic subtype were 9.69 years, 59%, and 49%, respectively, for 1692 synovial sarcoma patients in the United States who were diagnosed between 1975 and 2016. The cases were retrieved from the Surveillance, Epidemiology, and End Results program [19].

Conclusion

Our case study is an unusual example of synovial sarcoma in the perineum. MRI (T2-weighted sequences) allows for the identification of the triple sign in patients with synovial sarcoma. To distinguish synovial sarcoma from other sarcoma subtypes and to determine the tumor grade, biopsy, and histopathological evaluation are necessary. A multidisciplinary approach and early referral could improve care and outcomes.

Patient consent

Informed consent for patient information to be published in this article was obtained.

Author's contributions

Ho XT, Tran ND and Dau QL: Case file retrieval and case summary preparation. Dau QL and Nguyen MD: preparation of manuscript and editing. All authors read and approved the final manuscript.

Availability of data and materials

Data and materials used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Ethics approval and consent to participate

Our institution does not require ethical approval for reporting individual cases or case series. Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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