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A Rare Case of Peritoneal Synovial Sarcoma: A Case Report and Literature Review

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ABSTRACT

Synovial sarcoma is a malignant soft tissue tumor which commonly occurs in the young adult in the extremities. A few cases of the primary synovial sarcoma have been reported in the uncommon sites in the world literature. We report a case of primary synovial sarcoma arising from the peritoneum. This was about a 47-year old male who presented with abdominal fullness and pain. Abdominal ultrasound revealed a large hypoechoic tumor which was confirmed by computer tomography.

Surgery was performed. The tumor originated from the peritoneum, the morphology and immunochemical stains confirmed the diagnosis of biphasic synovial sarcoma. We represent the first case of primary synovial sarcoma arising from the peritoneum.

Keywords: Synovial sarcoma; Abdominal fullness; Peritoneum; Immunohistochemical stains

INTRODUCTION

Synovial sarcoma is a subtype of soft tissue malignant tumor, often occurring in the vicinity of a large joint; however, it has also been reported in a large variety of many uncommon sites, such as; abdominal wall, head and neck, retroperitoneum, mediastinum, cervix, fallopian tube, lung and maxillary sinus [1-9]. It is a rare soft tissue tumor, and mainly affects patients in their third decade [10]. Recently, we encountered a patient who developed synovial sarcoma from the peritoneum. Herein, we report this rare case to share our experience.

CASE DESCRIPTION

This 47-year old man visited a local gastrointestinal clinic because of abdominal fullness and pain for several days. Gastroscopic examination was performed; no tumor was found. Abdominal ultrasound study revealed a very large hypoechoic tumor, measuring 12.5x11.4 cm (Figure 1A & B). The patient was referred to a surgeon at the medical center. Labatory data including chemistry profile complete blood count were within normal limits. Computer tomography of the abdomen revealed a large heterogenous mass (Figure 2 A). Because of the size of the tumor, an open surgery was planned. After the surgeon opened the abdomen, a large tumor was found adhering to the mesocolon which could be easily separated (Figure 3 A). The tumor apparently originated from the peritoneum and loosely adhered to the internal oblique abdominal muscle. Adhesions were detached, and the peritoneal tumor was removed on bloc (Figure 3 B).

The tumor was well delineated and showed a fibrous pseudocapsule. The cut surface was multiloculated, with grayish tan nodules. Histologically, the tumor showed two components; spindle cells and clusters of epithelial cells, forming nests or glandular structures (Figure 4A & B). In immunohistochemistry, the tumor cells expressed strong nuclear staining with TLE1, strong epithelial membrane antigen, and cytokertin. CD 99 was weakly positive with cytoplasmic staining in focal areas (Figure 5A, B, C &D). Based on the characteristic biphasic pattern in morphology and immunohistochemical features, the diagnosis of synovial sarcoma was rendered. The tumor had 14 mitoses per 10 high power field, and necrotic area was <50%. According to FNCLCC Grade System [11], the final diagnosis was low grade biphasic synovial sarcoma. Eighteen days after surgery, computer tomogram displayed no residual tumor or metastasis (Figure 2B), No adjuvant chemotherapy or radiation therapy would be given. The patient would be followed closely.

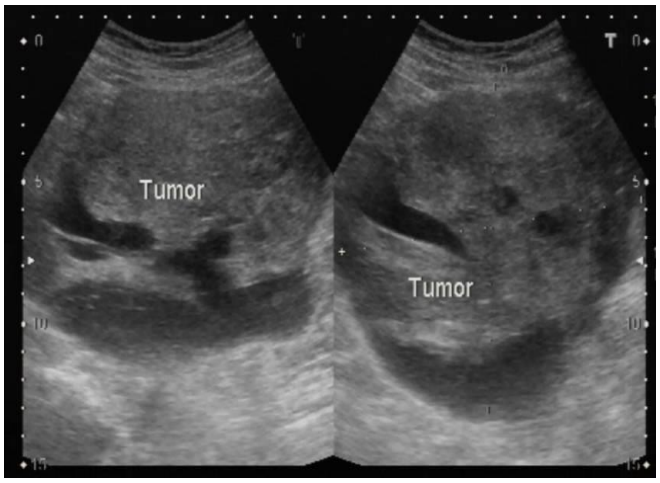

Figure 1A, B: Ultrasonography revealed a hypoechoic heterogenous tumor in the abdomen.

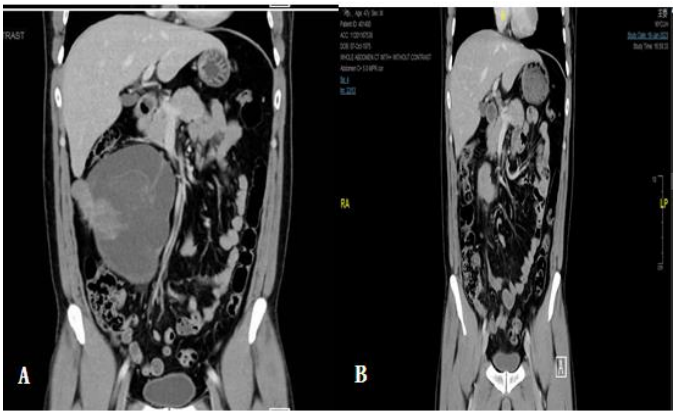

Figure 2: A. Computer tomography before surgery showed a large mass in the abdomen.

B. Computer tomography showed no residual tumor, 18 days after surgery.

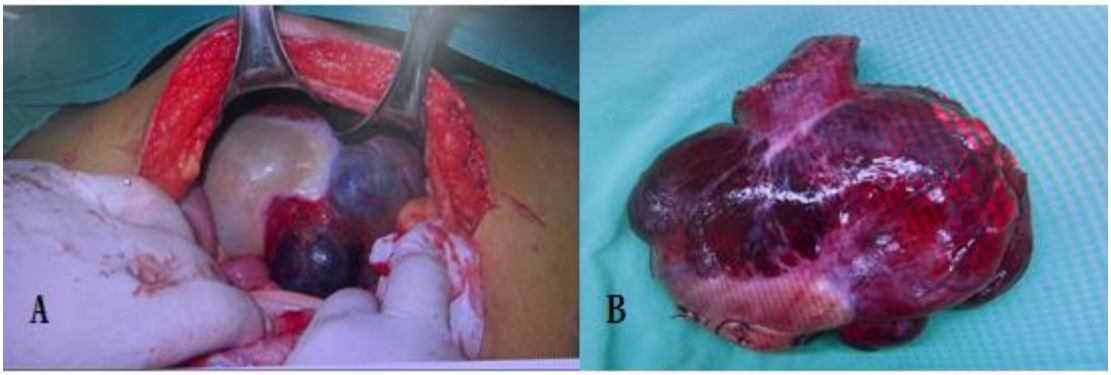

Figure 3: A. At surgery, a large tumor was found in the abdomen arising from the peritoneum, and adhered to the mesocolon.

B. the tumor was well delineated and showed a fibrous pseudocapsule, measuring 13x11x6 cm. Its cut surface was multiloculated, cystic with areas of grayish tan nodules.

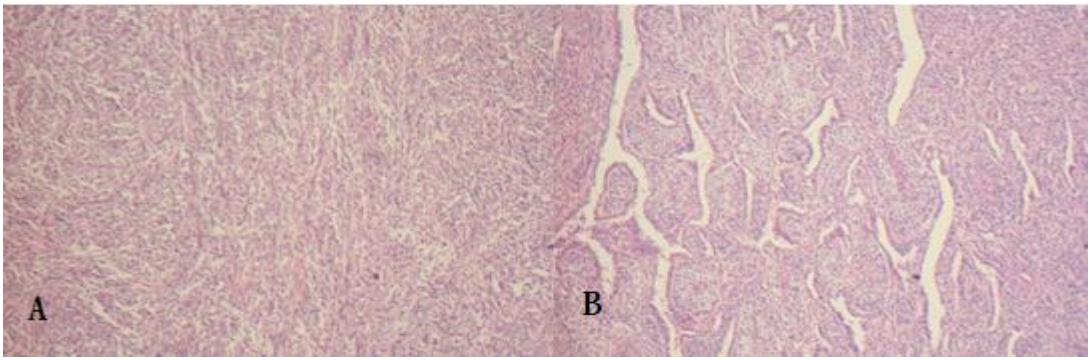

Figure 4: A. Spindle cells in the tumor, arranged in dense cellular sheets or vague fascicles. (H & E stain, x 200).

B. Tumor cells with glandular formation. (H & E stain, x 200).

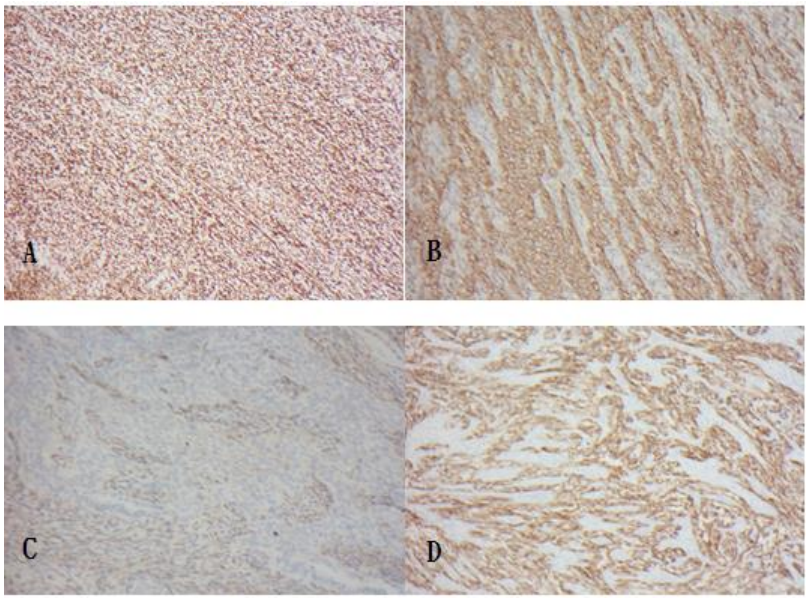

Figure 5: A. Diffuse and strong TLE1 nuclear staining, (Immunohistochemistry, x 200)

B. Diffuse and strong EMA expression, (Immunohistochemistry, x 200).

C. Focal weak CD 99 expression, (Immunohistochemistry, x 200).

D. Diffuse strong CK expression , (Immunohistochemistry, x 200).

DISCUSSION

Histologically, synovial sarcoma can be classified into biphasic, monophasic and poorly differentiated 3 subtypes . The biphasic type is characterized by the presence of a distinct epithelial component with glandular formation, and a distinct spindle cell component. The monophasic type consists of purely spindle cells. Poorly differentiated areas may be composed of fascicular spindle cells, small round cells or epithelioid cells [12]. Histological grade is the most important prognostic factor and the best indicator of metastatic risk. Therefore, it should be included in the pathology report. However, there is no specific grading system for synovial sarcoma. The majority of the pathologists applied the French grading and the National Cancer Institute grading which is used on the whole group on soft tissue sarcomas as a single entity [13, 14]. We hope that a specific grading system will become available in the near future.

Miettinen and Virtanen did a study on histogenesis of synovial sarcoma [15]. They used immunostains on normal synovial lining cells from the knee joint, and compared that with synovial sarcomas cells. They found out that the synovial

lining cells were not reactive with antibodies to keratin. Epithelial membrane antigen-like immunoreativity could not be detected in these cells either. These results were in contrast to that with synovial sarcoma cells. In addition, the lectin binding pattern were distinctly different between synovial lining cells and synovial sarcoma cells. They claimed that “synovial sarcoma” was a misnomer, and proposed a new term, such as” primary carcinosarcoma of blastoma of soft tissues” which took into account the ductal epithelial and mesenchymal differentiation. Then comes

the question where is the origin of synovial sarcoma? It is generally accepted that synovial sarcoma is originated from multipotent stem cells that are capable of differentiating into mesenchymal and/or epithelial structures [16]. This can explain why they occurred in so many unusual sites as we mentioned earlier in the introduction.

Genetically, synovial sarcoma harbors a unique t(X: 18) (p11, q 11) translocation [17].

By which SS18 on chromosome 18 is fused to one of the SSX genes (SSX1,SSX2,SSX3) on the X chromosome)[18].

In immunohistochemistry, the majority of synovial sarcoma are positive for CD 99, EMA, cytokeratin and BCL2 [19]. TLE1 is also positive for synovial sarcoma, but it is less specific [20]. To make the diagnosis of synovial sarcoma, the morphology and immunohistochemical stains are important. Cytogenetic study is recommended only in equivocal cases.

A wide surgical resection with safety margin is the standard care of choice. However, the recurrent rate ranged from 28-36% [1], close follow-up of at least 3 months for the first two years, and then every 6 months for another five years is recommended.

It is sometimes difficult to completely resecting tumor with safety margin in the locations such as head and neck, maxillary sinus or retroperitoneum, adjuvant chemotherapy or neoadjuvant chemotherapy can be used [3,9]. The benefit of chemotherapy in synovial sarcoma to overall survival remains unclear, although a study with advanced, poorly differentiated disease marginally improves with doxorubicin and ifosfamide treatment. The benefit of radiotherapy in synovial sarcoma is less clear than for chemotherapy [21].

CONCLUSION

Primary synovial sarcoma arising from the uncommon sites are rare, and has been reported in many occasions. Our case of primary synovial sarcoma arising from the peritoneum may represent the first case report. It is a misnomer to call them" synovial sarcoma", since the reported cases were all in extra-articular sites. Study in the literature has proved that synovial sarcoma occurring from the uncommon sites were arising from the primitive mesenchymal cells, rather than the synovial lining cells. Although complete removal of the tumor with the safety margin remains the standard of care, adjunctive radiation treatment and chemotherapy, either alone or in combination may reduce local recurrence and control metastasis, particularly in the sites complete removal are difficult. Close follow-up of the patients is mandatory.

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REFRENCES

1. Saif AH. Primary synovial sarcoma of the abdominal wall: a case report and review of the literature. J Family Community Med 2008;15(3):123-125.
2. Gopalakrishnan V, Amini B, Wagner MJ, Nowell EN, Lazar AJ,et al. Synovial Sarcoma of the Head

and Neck: A Single Institution Review. Sarcoma 2017;2016732

3. Wong CS, Harris A, Kennedy R, Houghton OP, Carey PD. A rare case of retroperitoneal synovial sarcoma. J of Royal Society of Med 2018;9(4):1-4
4. Ershadi R, Rahim M, Davari H. Primary mediastinal synovial sarcoma: A rare case report. Int J of Surg case report 2016:27(27):169-171.
5. Fard EV, Songlin Zhang, El Acho H, Covinsky M. Primary synovial sarcoma of the uterine cervix: First case report. Int J gynecological pathol 2021:40(2):196-203
6. Mitsuhashi A, Nagai Y, Yamazawa K, Nojima T, Nikaido T, et al. Primary synovial sarcoma in fallopian tube. Case report and literature review. Int J of gynecological pathol 2007:26(1):34-37.
7. Rajeev LK, Patidar R, Babu G, Babu MC, Lokesh KN, et al. A rare case of primary synovial sarcoma of lung. Lung India 2017:34(6):545-547.
8. Erich G, Hndan Z, Marian F, Thomas C, William T. Biphasic synovial saroma arising in the pleural cavity. A clinicalpathological study of five cases. Am J Surg Pathol 1996;20(1):36-45.
9. Saito S, Ozawa H, Ikari Y, Nakahara N, et al. Synovial sarcoma of the maxillary sinus: an extremely rare case with excellent response to chemotherapy. Oncotarget and therapy 2018;11:483-488
10. Eilber FC, Dry SM. Diagnosis and management of synovial sarcoma. J Surg Oncol 2008;97(4):314-320.
11. Coindre JM. Grading of soft tissue sarcoma: Review and Update. Arch Pathol Lab M 2006;(130):1448-1453.
12. Chan JA, McMeamin ME, Fletcher CD. Synovial sarcoma in older patients:Clinicopathological analysis of 32 cases with emphasis on unusual histological features. Histology 2003;43(1):72-83.
13. Brown FM, Fletcher CDM. Problems in grading soft tissue sarcoma. Am J ClinPathol 2000(114):582-589.
14. Evans HL. Classification and grading of soft tissue tumor. Hematol Oncol Clin North Am 1995;9:653-656.
15. Miettinen M, Virtanen I. Synovial sarcoma-A misnomer. Am J Pathol 1984:117(1):18-25.
16. Skytting B. Synovial sarcoma. A Sandinavian Sarcoma Group project. Acta Orthop Scand 2000;291:SI-S28.
17. Ladanyi M, Antonescu CR, Leung DH, et al. Impact of XYT-SSX fusion type on the clinical behavior of synovial sarcoma: a multi-institutional retrospective study of 243 patients. Cancer Res 2002;62(1):135-140.
18. Santos D, Bruijn DR, Kessel AG. Molecular mechanism underlying human synovial sarcoma development Genes Chromosome Cancer 2001;30(1):1-14.
19. Chen SH, Thomas DG, Lucas DRC. Cluster analysis of immunohistochemical profile on synovial sarcoma, malignant peripheral nerve sheath tumor, Ewing sarcoma. Modern Histology 2006:19(5):659-668.
20. Foo WC, Cruise MW, Wick MR, et al. Immunohistochemical staining for TLE1distinguishs synovial

sarcoma from histological mimics. Am J Clin Pathol 2011:135(6):839-844.

21. Thway K, Fisher C. Synovial sacoma defining features and diagnostic evolution. Ann Diagn Pathol 2014; 18:369-380.