Acta Scientific Gastrointestinal Disorders (ISSN: 2582-1091)

Volume 4 Issue 10 October 2021

Case Report

Synovial Sarcoma of the Liver - A Case Report

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Received: August 27, 2021

Published: September 23, 2021

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Abstract

Synovial sarcoma is an uncommon malignant mesenchymal neoplasm that usually occurs in extremities. Rarely have they been reported in the head and neck, pericardium, lungs, liver, and retroperitoneum. The term synovial sarcoma is a misnomer because the tumor doesn't arise from synovial tissue. It was once believed to recapitulate synovium, but the cell of origin is still unclear. We report a case of synovial sarcoma of the liver in a 55-year-old male, presented to our hospital with a history of abdominal discomfort and weight loss.

Keywords: Liver; Synovial Sarcoma; Translocation; Immunohistochemistry; Monophasic; Biphasic

Abbreviations

USS: Ultrasound Scan; CT: Computed Tomography; FISH: Fluorescence *In Situ* Hybridization; RT-PCR: Reverse Transcription-Polymerase Chain Reaction

Introduction

Among the soft tissue sarcoma in adults, the third most common is synovial sarcoma. It accounts for approximately 10% of soft-tissue sarcomas. Although most synovial sarcomas occur in the lower extremities, rare cases originate in the head and neck, thorax, and abdomen [1]. Synovial sarcoma is thought to arise from primitive mesenchymal cells that undergo differentiation to resemble synovial cells. The liver, the largest solid organ in the body, has a propensity to develop common primary hepatic neoplasms like hepatocellular carcinoma and cholangiocarcinoma and harbor metastatic secondary deposits from primary neoplasm elsewhere. But, though less common, the tumors that arise from the hepatic mesenchyme and capsule-like sarcoma merit consideration. As the imaging features of this rare sarcoma are non-specific, histopathological, immunohistochemistry, and cytogenetic examinations are crucial for diagnosis. The specific translocations for synovial sarcoma are t(X;18)(p11.2;q11.2) and the chimeric gene SYTSSX.

Case Report

A 55-year-old man presented with complaints of weight loss and abdominal discomfort. The initial ultrasound scan (USS) showed multiple lesions in both lobes of the liver, with the dominant tumor in the left lobe of the liver. Contrast-enhanced computed tomography (CT) of the abdomen was performed, which showed multiple heterogeneously enhancing lesions of varying sizes in both lobes of the liver. The lesions showed mild enhancement on the arterial phase and progressive filling in the venous phase with central non-enhancing necrotic areas. The largest lesion was noted in the left lobe of the liver (Figure 1). The lesion mentioned above also infiltrated the greater curvature of the stomach (Figure 2) and the adjacent omentum. Few lung nodules were also noted in the

thorax (Figure 3). The possibility of cholangiocarcinoma with liver and lung metastasis was given. The patient underwent USG guided biopsy from the mass lesion in the left lobe of the liver, and the samples were sent for both histopathologic examination and immunohistochemistry.

Figure 1

Figure 2

Figure 3

The biopsy was positive for malignancy and showed hyperchromatic spindled cells (Figure 4) and the possibility of spindle cell malignancy was raised. On immunohistochemistry analysis, the following markers were positive: Vimentin, CD 99, TLE 1, and Pan CK. The following markers were negative; Desmin, S100, CD 34, CD117, EMA, Hep-Par 1. IHC with histology favored the diagnosis of synovial sarcoma - a poorly differentiated type. Though identification of the translocation of chromosome (X; 18) (p11.1; q11.2) is specific for the diagnosis of synovial sarcoma, the patient refused to undergo the study. The patient also had hepatic and renal decompensation. Because of extensive disease and associated co-morbidities, the patient was declared medically unfit for sarcoma-directed therapy. He was managed symptomatically, and the patient died within two months.

Figure 4

Discussion

The hepatic sarcomas include angiosarcomas, epithelioid hemangioendothelioma, undifferentiated embryonal sarcoma, and rhabdomyosarcoma. Fibrosarcomas, malignant fibrous histiocy-

toma, and leiomyosarcomas are even more unusual. Synovial sarcoma occurs mainly in young adults and teenagers, although they also occur in older adults and preteen children.

The synovial sarcoma most commonly occurs in the lower limbs. The synovial sarcoma arises from the lower extremities approximately in 70% of cases [1]. The liver is an infrequent location for this tumor type. Therefore, the possibility of a secondary synovial sarcoma should always be excluded. Multiple hepatic lesions were observed in the present case, which was suggestive of metastatic disease. It is doubtful for a synovial sarcoma to present as an occult primary.

In the present case, imaging studies and physical examinations ruled out the possibility of primary tumor elsewhere. In synovial sarcoma, the main histologic variants described are classic biphasic type (spindle cells and epithelial cells), monophasic type (made only spindle cells), and a poorly differentiated type (cells that resemble those of small round blue cell tumors) [3]. In our case, spindle cell neoplasms with muscle origin and malignant peripheral nerve sheath tumor were excluded by desmin and S- 100 negativity of tumor cells, respectively. Most of the synovial sarcomas, about 90%, express cytokeratins in the epithelial component and in some clusters of spindle cells. Vimentin is expressed in the spindle cell component. Some of them may express CD99 and S-100 protein [4]. Since the imaging findings of these synovial sarcomas are nonspecific, immunohistochemistry and cytogenetic analysis play a significant role in its diagnosis.

The hallmarks for synovial sarcoma are the t(X;18) translocation and SYT-SSX gene fusion products. These can be identified using FISH (fluorescence *in situ* hybridization) or RT-PCR (reverse transcription-polymerase chain reaction) studies, respectively. This genetic aberration has been noted in greater than 90% of synovial sarcomas and is highly specific [2].

Conclusion

In conclusion, though it is rare, the tumors arising from the liver mesenchyme and the capsule like synovial sarcoma should also be considered in differential diagnosis for a heterogeneously enhancing liver lesion.

Conflict of Interest

Nil.

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Volume 4 Issue 10 October 2021
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