Primary Hepatic Synovial Sarcoma with Synchronous Colorectal Adenocarcinoma: A Case Report and Review of Literature

Xuan Vinh Vu

Bach Mai Hospital

Thanh Khiem Nguyen

Bach Mai Hospital

Ham Hoi Nguyen

Bach Mai Hospital

Ngoc Hung Nguyen

Bach Mai Hospital

Tuan Hiep Luong (✉ hiep1995hsgs@gmail.com)

Hanoi Medical University

Cong Nguyen Nguyen

Bach Mai Hospital

Van Chuong Tran

Bach Mai Hospital

Case Report

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Abstract

Background: Primary hepatic synovial sarcoma is an extremely rare disease with a poor prognosis. The symptoms are nonspecific, and it is usually diagnosed at an advanced stage.

Case presentation: A 63-year-old male patient was admitted with asymptomatic but elevated Alpha Fetoprotein (AFP) serum level in routine medical examination with a medical history of Hepatitis B with Tenofovir 300 mg per day. No sign of jaundice was observed. Her abdomen was flat and soft without tenderness or palpable mass. Dynamic abdominal computed tomography (CT) scan and magnetic resonance imaging (MRI) showed a hypervascular 15 x 26 mm mass within hepatic subsegment V. There was no distant spread location. Colonoscopy revealed 3-cm sessile polyp in Sigmoid colon with pathological result was adenocarcinoma. Immunohistochemistry was performed to accurate diagnosis. The tumor cells were positive for CD99, EMA and a Ki67 value of 40% and were negative for Hepar-1, CD34, CD117, CK7, STAT6, DOG1, SMA and S100. The final diagnosis for this patient was a Malignant Sigmoid-colon polyp with a Primary hepatic synovial sarcoma. Laparoscopic Sigmoid-colon and rectal resection with injecting autologous blood to mark the tumor before surgery was performed for malignant polypectomy lesion and Hepatic Radiofrequency Ablation (RFA) was performed four days before surgery. With no postoperative complications as well as acute hepatic failure, the patient was discharged on the POD8 uneventfully.

Conclusion: A primary hepatic synovial sarcoma is especially rare, with the present case being the fifth accounted in the English literature. Synovial sarcoma (SS) is a genetically soft tissue sarcoma with uncertain differentiation and originated from primitive mesenchymal cells. We here present a report of this case with had primary hepatic synovial sarcoma synchronously with a colorectal adenocarcinoma.

1. Background

Synovial sarcomas (SS) are rare, aggressive sarcomas of primitive mesenchymal cells, set in a hemangiopericytoma (HPC)-like vascular pattern, which account for 5%-10% of all soft tissue sarcoma [1, 2]. Synovial sarcomas can be found in any part of the body, primarily arising in the extremities, especially in the anatomic structures adjacent to the knee joint, but extremely rare in the abdominal viscera, especially the liver, with only a few cases reported in the literature [3–7]. The specific etiology of synovial sarcoma remains unknown. Several risk factors have been associated with poor prognosis, including tumor size, anatomic visceral sites as well as low-graded [8–10]. The clinical signs and symptoms of hepatic synovial sarcoma are nonspecific. Survival outcome was occasionally poor, with the five-year survival rate is about 50–60% [11–13]. We herein present a report of a primary hepatic synovial sarcoma with a Malignant Sigmoid colon polyp. This report can be useful for both education and clinical practice purposes. All our work has been reported in line with the CARE criteria and guidelines [14].

2. Case Presentation

A 63-year-old male patient was admitted with asymptomatic but elevated Alpha Fetoprotein (AFP) serum level in routine medical examination. He had a medical history of Hepatitis B with Tenofovir 300 mg per day. He denied current or prior alcohol, smoking or with no known exposure to carcinogens and drugs. Physical examination showed clinical signs within normal limits. No sign of jaundice was observed. Her abdomen was flat and soft without tenderness or palpable mass. The AFP serum level was 43.5 ng/ml. Dynamic abdominal computed tomography (CT) scan showed a heterogenous 15 x 26 mm mass within hepatic subsegments IV-V. Similarly, Magnetic resonance imaging (MRI) showed a heterogenous mass with low and high intensity on T1 and T2-weighted images with post-gadolinium enhancement, respectively. There was no distant spread location (Fig. 1). Colonoscopy revealed 3-cm sessile polyp in Sigmoid colon with pathological result was adenocarcinoma. A percutaneous liver biopsy fine needle aspirations (FNA) was proceeded, and the result was Spindle cell tumor. Immunohistochemistry was performed to accurate diagnosis. The tumor cells were positive for CD99, EMA and a Ki67 value of 40% and were negative for Hepar-1, CD34, CD117, CK7, STAT6, DOG1, SMA and S100 (Fig. 2). The final pathological result was a Hepatic primary synovial sarcoma. The final diagnosis for this patient was a Malignant sigmoid-colon polyp with a Primary hepatic synovial sarcoma. MDT meeting consisted of surgeons, physicians, clinical and medical oncologists, radiologists, pathologists, and clinical nurse specialists (CNSs) were organized to make clinical decisions. The informed consent was signed. Laparoscopic Sigmoid-colon and rectal resection with injecting autologous blood to mark the tumor before surgery was performed for malignant polypectomy lesion. Four days before surgery, Hepatic Radiofrequency Ablation (RFA) was performed for Hepatic primary synovial sarcoma. With no postoperative complications as well as acute hepatic failure, the patient was discharged on the POD8 uneventfully. At the most recent 6 months follow-up after the operation, he was still alive without local recurrence and distant metastases were identified.

3. Discussion And Conclusion

Synovial sarcoma (SS) is a genetically soft tissue sarcoma with uncertain differentiation, with originate from primitive mesenchymal cells, not from intra-articular synovium like its misnomer [1, 13]. Despite of being a rare malignancy sarcoma with an incidence of 800 to 1,000 cases per year in the United States, adolescents and adults younger than 30 years of age were believed the most common but favorable prognostic group of patients [15]. In location, most primary tumors were found near articular structures, especially in the lower extremities [1]. However, primary synovial sarcomas which was raised from uncommon locations such as mediastinum, retroperitoneum, and various viscera has been reported [1, 2, 13]. To our best knowledge, there were only 4 cases of primary hepatic synovial sarcoma which have been reported in the English-language literature [3–6]. The age of the 4 reported cases was 13, 18, 44, and 60 years, respectively. So that, with a case of hepatic spindle cell tumors was found, the possibility of metastases from extrahepatic sarcomas should always be excluded [3]. In our case, all imaging and physical examinations have excluded the possibility of secondary tumor.

In imaging characteristics, liked all other soft-tissue sarcomas, magnetic resonance imaging (MRI) is the gold standard for diagnostic imaging, with typically well-defined and heterogenous solid mass with low

and high intensity on T1 and T2-weighted images with post-gadolinium enhancement, respectively [16]. Computed tomography (CT) with contrast can be applied for cases with untypical results in MRI. With cases of liver metastasis, on CT, SS had hypodense appearance comparing with muscle as well as heterogeneous enhancement with calcifications [17]. In our cases, all MRI and CT were used, and the tumor has appeared with similar appearances. Due to the patient's medical history of Hepatitis B and elevated Alpha Fetoprotein (AFP) serum level in routine medical examination, the most-likely diagnosis was Hepatocellular carcinoma (HCC), but it hadn't met the criteria of the 2018 ESMO Clinical Practice Guidelines for diagnosis in Imaging characteristics and Alpha Fetoprotein (AFP) serum level [18], so that, we have proceeded to take a percutaneous liver biopsy with fine needle aspirations (FNA) and pathologic assessment for accurate diagnosis.

Microscopically and histologically, all the 5 cases (including the current one) exhibited classic features of synovial sarcoma with spindle cells. That represents for the monomorphic appearance, and other two sub-types were biphasic (associated with variable epithelial differentiation) or poorly differentiated (with greater nuclear atypia and high mitotic activity) [1]. On immunohistochemistry, about 60% of all cases, tumoral cells were positive for CD99, as well as always negative for CD34 [19]. In our case, the tumor stain positive focally for Epithelial Membrane Antigen (EMA), represented epithelial differentiation in this case. So that, we have consumed that our tumor was biphasic sub-type. In molecular pathology, a pathognomonic translocation between chromosomes X and 18, or $t(X;18)(p11.2;q11.2)$, was characterized for SS, with an rate of over 95% of cases, with can be identified by exact fusion transcript (*SS18-SS X7*) [13, 20]. Due to its rarity, the differential diagnosis of primary hepatic synovial sarcoma is quite broad, with other more common types of primary liver sarcomas, such as hemangioendothelioma, angiosarcoma, leiomyosarcoma, embryonal rhabdomyosarcoma, carcinosarcoma and giant cell sarcoma [21]. In our case, the tumor stain negative for Hepar-1, DOG1, CD117, STAT6, CD34, SMA and S100. So that, we can occasionally rule out the possibility of hepatocellular carcinoma, gastrointestinal stromal tumor, other types of solitary fibrous tumors [22–24].

And due to the rarity of primary liver sarcomas (PLS), especially primary hepatic SS, the optimal treatment approach is still not well-defined, which was depended on histological type and the ability to accomplish negative resection margins[21, 25]. In our case, with tumoral position deeply within hepatic subsegments IV-V and near important anatomical landmarks that are right and middle hepatic veins, as well as small size (about 2cm), we had decided to proceed Hepatic Radiofrequency Ablation (RFA), due to its utility of local tumor control with minimal morbidity in patients with small-sized hepatic tumors compared to liver resection [26]. However, with high-rated of local recurrence and distant metastasis, especially in elderly populations and high Ki-67 index, primary synovial sarcoma must be follow-up carefully for early detection and management [1].

Conclusion

To the best of our knowledge, the present patient is the fifth case report of primary hepatic synovial sarcoma. Imaging features are non-specific, with the diagnosis being only established through histology

and immunohistochemistry analysis. Our discussion emphasizes the difficulty associated with diagnosis and management of this rare hepatic neoplasm.

Abbreviations

CD: Cluster of differentiation; CK: Cyclin-dependent kinase; CT: Computed tomography; DOG1: Discovered on GIST-1; MRI: Magnetic resonance imaging; RFA: Radiofrequency Ablation; STAT6: Signal transducer and activator of transcription 6, EMA: Epithelial membrane antigen; SMA: Smooth muscle actin.

Declarations

Ethics approval and consent to participate

Ethics approval of this study was given by the Research Ethics Committees of Bach Mai Hospital. Authors have agreed to submit it in its current form for consideration for publication in the Journal.

Consent for publication

Written informed consent for publication of their clinical details and clinical images was obtained from the patient's next of kin.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Competing interests

The authors declare that there is no conflict of interest regarding the publication of this article.

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Authors' contributions

XVV and TKN contributed equally as co-first authors, the main doctors conceived the original idea and operated the patients; HHN and NHN edited the manuscript; THL performed the operation and wrote the manuscript; CNN performed the operation; VCT edited the manuscript. The co-first authors and the other authors discussed the results together and contributed to the final manuscript. All authors read and approved the final manuscript.

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Author details

¹ Department of Gastrointestinal and Hepato-pancreato-biliary surgery, Bach Mai Hospital, Hanoi, Vietnam. ² Department of Surgery, Hanoi Medical University, Vietnam. ³ Pathology Center, Bach Mai Hospital, Hanoi, Vietnam

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Figures

Figure 1

(A) Dynamic abdominal computed tomography (CT) scan showed a heterogenous 15 x 26 mm mass within hepatic subsegment IV-V.

(B) Magnetic resonance imaging (MRI) showed a heterogenous mass with low intensity on T1 and high intensity on T2-weighted images with post-gadolinium enhancement.

Figure 2

(A) (B) The result of hematoxylin and eosin (H&E) and Periodic acid Schiff (PAS) staining was Spindle cell tumor.

(C) (D) (E) Immunohistochemical results were positive for CD99, EMA (focally) and a Ki67 value of 40%, respectively.