

World Journal of *Gastroenterology*

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EDITORIAL

- 2269 Gastroesophageal reflux disease and morbid obesity: To sleeve or not to sleeve?
Rebecchi F, Allaix ME, Patti MG, Schlottmann F, Morino M

REVIEW

- 2276 Advanced pancreatic ductal adenocarcinoma - Complexities of treatment and emerging therapeutic options
Divakarla C, Hamman K, Hein N, Yip D

MINIREVIEWS

- 2286 Indoleamine 2,3-dioxygenase: As a potential prognostic marker and immunotherapeutic target for hepatocellular carcinoma
Asgar K, Farooq A, Zulfiqar B, Rashid MU

ORIGINAL ARTICLE

Basic Study

- 2294 Disruption of the TWEAK/Fn14 pathway prevents 5-fluorouracil-induced diarrhea in mice
Sezaki T, Hirata Y, Hagiwara T, Kawamura YI, Okamura T, Takanashi R, Nakano K, Tamura-Nakano M, Burkly LC, Dohi T
- 2308 CMA down-regulates p53 expression through degradation of HMGB1 protein to inhibit irradiation-triggered apoptosis in hepatocellular carcinoma
Wu JH, Guo JP, Shi J, Wang H, Li LL, Guo B, Liu DX, Cao Q, Yuan ZY
- 2318 Cullin 4A is associated with epithelial to mesenchymal transition and poor prognosis in perihilar cholangiocarcinoma
Zhang TJ, Xue D, Zhang CD, Zhang ZD, Liu QR, Wang JQ
- 2330 Notch signaling mediated by TGF- β /Smad pathway in concanavalin A-induced liver fibrosis in rats
Wang Y, Shen RW, Han B, Li Z, Xiong L, Zhang FY, Cong BB, Zhang B
- 2337 MicroRNA-145 exerts tumor-suppressive and chemo-resistance lowering effects by targeting CD44 in gastric cancer
Zeng JF, Ma XQ, Wang LP, Wang W
- #### Case Control Study
- 2346 Predictors for difficult cecal insertion in colonoscopy: The impact of obesity indices
Moon SY, Kim BC, Sohn DK, Han KS, Kim B, Hong CW, Park BJ, Ryu KH, Nam JH

Retrospective Cohort Study

- 2355** Impact of interferon-free antiviral therapy on lipid profiles in patients with chronic hepatitis C genotype 1b
Endo D, Satoh K, Shimada N, Hokari A, Aizawa Y

Retrospective Study

- 2365** Transition after pediatric liver transplantation - Perceptions of adults, adolescents and parents
Junge N, Migal K, Goldschmidt I, Baumann U
- 2376** Minimally invasive surgery for gastric cancer: A comparison between robotic, laparoscopic and open surgery
Parisi A, Reim D, Borghi F, Nguyen NT, Qi F, Coratti A, Cianchi F, Cesari M, Bazzocchi F, Alimoglu O, Gagnière J, Pernazza G, D'Imporzano S, Zhou YB, Azagra JS, Facy O, Brower ST, Jiang ZW, Zang L, Isik A, Gemini A, Trastulli S, Novotny A, Marano A, Liu T, Annecchiarico M, Badii B, Arcuri G, Avanzolini A, Leblebici M, Pezet D, Cao SG, Goergen M, Zhang S, Palazzini G, D'Andrea V, Desiderio J
- 2385** Clinical implication of FDG uptake of bone marrow on PET/CT in gastric cancer patients with surgical resection
Lee JW, Lee MS, Chung IK, Son MW, Cho YS, Lee SM

Observational Study

- 2396** Safety and efficacy of tenofovir in chronic hepatitis B-related decompensated cirrhosis
Lee SK, Song MJ, Kim SH, Lee BS, Lee TH, Kang YW, Kim SB, Song IH, Chae HB, Ko SY, Lee JD
- 2404** Can mean platelet volume play a role in evaluating the severity of acute pancreatitis?
Lei JJ, Zhou L, Liu Q, Xiong C, Xu CF

Prospective Study

- 2414** Proposed criteria to differentiate heterogeneous eosinophilic gastrointestinal disorders of the esophagus, including eosinophilic esophageal myositis
Sato H, Nakajima N, Takahashi K, Hasegawa G, Mizuno K, Hashimoto S, Ikarashi S, Hayashi K, Honda Y, Yokoyama J, Sato Y, Terai S
- 2424** Therapeutic experience of 289 elderly patients with biliary diseases
Zhang ZM, Liu Z, Liu LM, Zhang C, Yu HW, Wan BJ, Deng H, Zhu MW, Liu ZX, Wei WP, Song MM, Zhao Y

META-ANALYSIS

- 2435** What is the quantitative risk of gastric cancer in the first-degree relatives of patients? A meta-analysis
Yaghoobi M, McNabb-Baltar J, Bijarchi R, Hunt RH

CASE REPORT

- 2443** Hepatic angiosarcoma with clinical and histological features of Kasabach-Merritt syndrome
Wadhwa S, Kim TH, Lin L, Kanel G, Saito T

LETTERS TO THE EDITOR

2448 Tumor biopsy and patient enrollment in clinical trials for advanced hepatocellular carcinoma

Rimassa L, Reig M, Abbadessa G, Peck-Radosavljevic M, Harris W, Zagonel V, Pastorelli D, Rota Caremoli E, Porta C, Damjanov N, Patel H, Daniele B, Lamar M, Schwartz B, Goldberg T, Santoro A, Bruix J

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Hepatic angiosarcoma with clinical and histological features of Kasabach-Merritt syndrome

Sanya Wadhwa, Tae Hun Kim, Leah Lin, Gary Kanel, Takeshi Saito

Sanya Wadhwa, Department of Medicine, Keck School of Medicine, University of Southern California, Los Angeles, CA 90033, United States

Takeshi Saito, Department of Medicine, Molecular Microbiology and Immunology, and Pathology, Division of Gastrointestinal and Liver Diseases, Keck School of Medicine of USC, University of Southern California, Los Angeles, CA 90033, United States

Tae Hun Kim, Gary Kanel, Department of Pathology, University of Southern California, Keck School of Medicine, Los Angeles, CA 90033, United States

Leah Lin, Department of Radiology, University of Southern California, Keck School of Medicine, Los Angeles, CA 90033, United States

Author contributions: Wadhwa S gathered information of the case, reviewed the literature, and wrote the manuscript; Kim TH and Kanel G provided pathological interpretation of liver biopsy; Lin L provided interpretation of radiographic imaging, Saito T oversaw the manuscript preparation; all of the authors contributed to the intellectual content.

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Correspondence to: Takeshi Saito, MD, PhD, Department of Medicine, Molecular Microbiology and Immunology, and Pathology, Division of Gastrointestinal and Liver Diseases, Keck School of Medicine of USC, University of Southern California, 2011 Zonal Avenue, HMR 801A, Los Angeles, CA 9003, United States. saitotak@usc.edu
Telephone: +1-323-4422260
Fax: +1-323-4425425

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Abstract

Hepatic angiosarcoma is a mesenchymal tumor originating from liver sinusoidal endothelial cells. It is an extremely rare malignant neoplasm accounting for less than 1% of primary malignant liver tumors. The deregulated coagulopathy that can be seen in hepatic angiosarcoma fulfills the clinical diagnostic criteria of disseminated intravascular coagulation. However, the mechanism that governs this coagulopathy has been poorly understood. This case report provides histological evidence of the consumption of coagulation factors along with trapped platelets occurring within the tumor, which is the foundation for the concept of Kasabach-Merritt syndrome (KMS). KMS is characterized by thrombocytopenia and hyperconsumption of coagulation factors within a vascular tumor. However, KMS associated with angiosarcoma has not been well recognized. This case report describes, for the first time, the histological evidence of KMS that occurred in an extremely rare mesenchymal malignant tumor of the liver.

Key words: Hepatic angiosarcoma; Kasabach-Merritt syndrome; Vascular tumor

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Core tip: Kasabach-Merritt syndrome (KMS) is characterized by thrombocytopenia and hyperconsumption of coagulation factors within a vascular tumor. KMS is typically seen in the pediatric population however there have been reports of KMS occurring in association with adult vascular tumors. Based on laboratory findings, it is hard to differentiate KMS from disseminated intravascular coagulation. Here, we describe, for the first time the histological evidence validating the concept of KMS.

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INTRODUCTION

Hepatic angiosarcoma arises from vascular endothelial cells within the liver^[1]. It is believed that in the past, the malignant transformation of vascular endothelial cells was mediated or triggered by environmental or industrial toxins, such as vinyl chloride, arsenic, and thorium dioxide, however despite tighter regulations of these toxins, there is still a constant number of reports of angiosarcoma without any such association

The clinical entity of hepatic angiosarcoma has been well known to have an extremely poor prognosis. The disease presentation typically manifests as abdominal distension from large hepatic mass effect or by its complication of tumor rupture into abdominal cavity or intra-tumor bleeding. Subsequently, the average lifespan after the diagnosis has been reported to be 6 mo^[2]. To date, there is no definitive or effective treatment that has been established. An attempt of surgical resection or liver transplant does not provide significant advantage in extending life as the tumor recurs in nearly all cases reported^[3]. This resulted in the median life expectancy of 16 and 5 mo in partial hepatectomy and liver transplant, respectively^[3,4]. In addition, there are no well-established chemotherapy regimens and accordingly, this has been primarily utilized as a palliative measure^[3].

The unique clinical characteristic of angiosarcoma is the pronounced dysregulation of the coagulation system seen in a subset of patients^[5,6]. This is featured by significant thrombocytopenia, prolonged prothrombin time (PT), and activated partial thromboplastin (aPTT) time with significant elevation of fibrin net degradation products, such as d-dimer. These abnormalities are congruent with the diagnostic criteria of disseminated intravascular coagulation (DIC). However, the site and

mechanism of the clinical feature of this abnormal coagulopathy associated with angiosarcoma have never been well explained. The clinical entity of Kasabach-Merritt syndrome (KMS) is indistinguishable from DIC by blood-based laboratory findings. The coagulopathy associated with KMS is characterized by hyper-activation of the coagulation cascade and entrapment of platelets within dilated sinusoids of the vascular tumor^[7]. The concept of KMS is well described in the pediatric population with Kaposi hemangioendothelioma, and to a much lesser extent seen in giant hemangioma in the adult population^[8]. Here we describe a case of primary hepatic angiosarcoma with the clinical and for the first time, histological evidence of KMS.

CASE REPORT

A 44-year-old Hispanic male was admitted to our hospital for worsening abdominal pain and jaundice. Abdominal ultrasound demonstrated multiple masses with heterogeneous echogenicity (Figure 1). Color doppler study revealed hypervascularity within the tumors.

The patient had no significant past medical, family, or social history. He had no prior exposures to vinyl chloride, arsenic, or thorium dioxide. The physical examination was significant for scleral icterus and a distended abdomen with diffuse tenderness. The exam was negative for spider angiomas, palmar erythema, shifting dullness, hepatic bruit, caput medusae, and asterixis.

Pertinent laboratory values on admission were as follows: white blood cell count 13400/mm³, hemoglobin 8.8 g/dL, mean corpuscular volume 99 fL, platelets 57000/mm³, alkaline phosphatase 118 IU/L, total protein 5.4 g/dL, albumin 2.7 g/dL, total bilirubin 6.4 mg/dL, direct bilirubin 3.1 mg/dL, aspartate aminotransferase 42 IU/L, alanine transaminase 72 IU/L, prothrombin time 29.4 s, INR 2.88, PTT 40.7 s, fibrinogen less than 60 mg/dL, and d-dimer greater than 9.999 mg/dL. Serologies for viral hepatitis and auto-immune liver disease were negative. Tumor markers such as AFP, CA 19-9, and CEA were all within normal limits.

Multiphase computerized tomography and magnetic resonance imaging of the abdomen revealed discrete, multifocal, and isodense masses in precontrast images involving all segments of the liver, with the largest measuring 6.5 cm (Figures 2 and 3). Peripheral enhancement was seen in the arterial phase, but not in portal and delayed phases. Of note, there was no definite washout. These images did not demonstrate features of commonly identified hepatic malignancies, such as hepatocellular carcinoma and intrahepatic cholangiocarcinoma. The patient subsequently underwent liver needle biopsy.

The tissue demonstrated a mesenchymal tumor infiltrating the sinusoids with anastomosing, dilated vascular channels lined by atypical cells (Figure 4A).

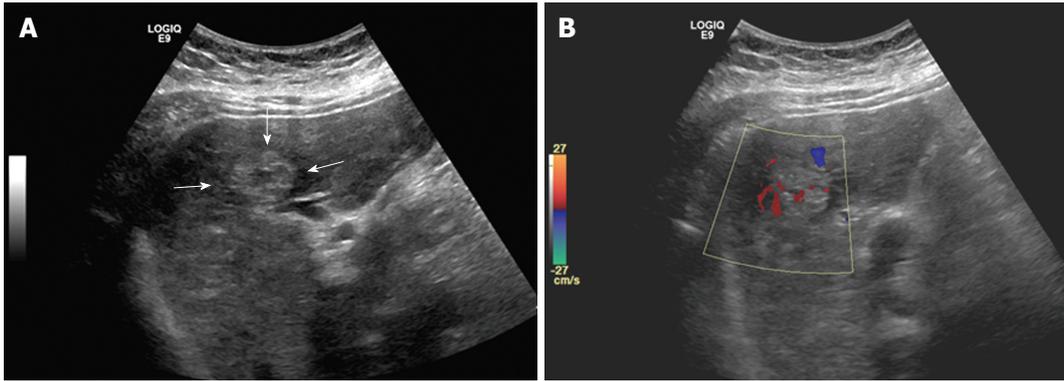


Figure 1 Ultrasonographic findings of hepatic angiosarcoma. A: Representative image of tumor detected by abdominal ultrasound (arrow). Note the increased peripheral echogenicity and hypoechoic central appearance; B: Color doppler study of (A) demonstrates hypervascularity of the tumor. No arterial flow was detected (image not shown).

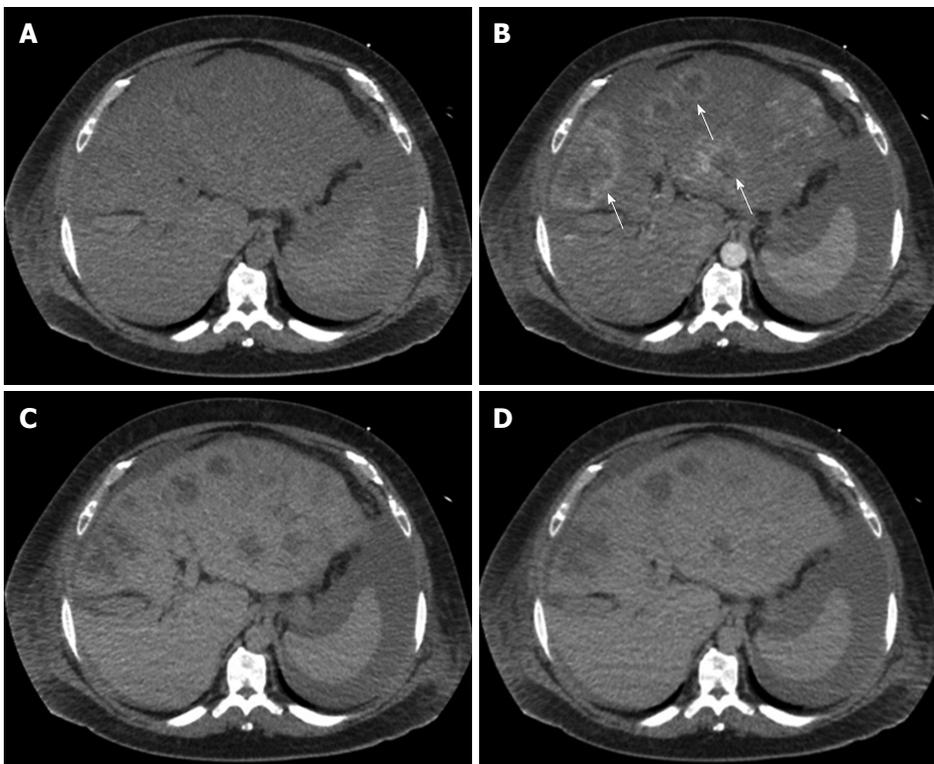


Figure 2 Multi-phase axial computerized tomography images of hepatic angiosarcoma. A: Precontrast image fails to visualize the tumor likely due to its isodensity; B: Arterial phase demonstrates discrete, multifocal masses with peripheral enhancement (arrow); C and D: Portal venous and delayed phase images show discrete masses without enhancement and without definite washout.

Immunohistochemical studies demonstrated tumor cells strongly expressed CD34, suggesting a vascular endothelial origin (Figure 4B). Of note, the uninvolved region showed no evidence of chronic liver disease. Based on these findings, the diagnosis of angiosarcoma was made.

DISCUSSION

In this case report we described a case of hepatic angiosarcoma in a young male with no evidence of cirrhosis and no prior history of exposure to the aforementioned chemicals through his occupation or

medical use. In addition, this patient had no history of use of anabolic steroids or conditions that were associated with the onset of angiosarcoma. Therefore, the diagnosis of idiopathic hepatic angiosarcoma was made. Consistent with other reports, this case also manifested with significant coagulopathy which is a unique feature of hepatic angiosarcoma. As described in our case, the results of the blood tests fit well into the diagnostic criteria of DIC.

Our histological investigation found increased expression of von-Willebrand factor (vWF)/factor VIII within the tumor cells, the formation of fibrin nets, and platelet aggregation within the dilated sinusoids of

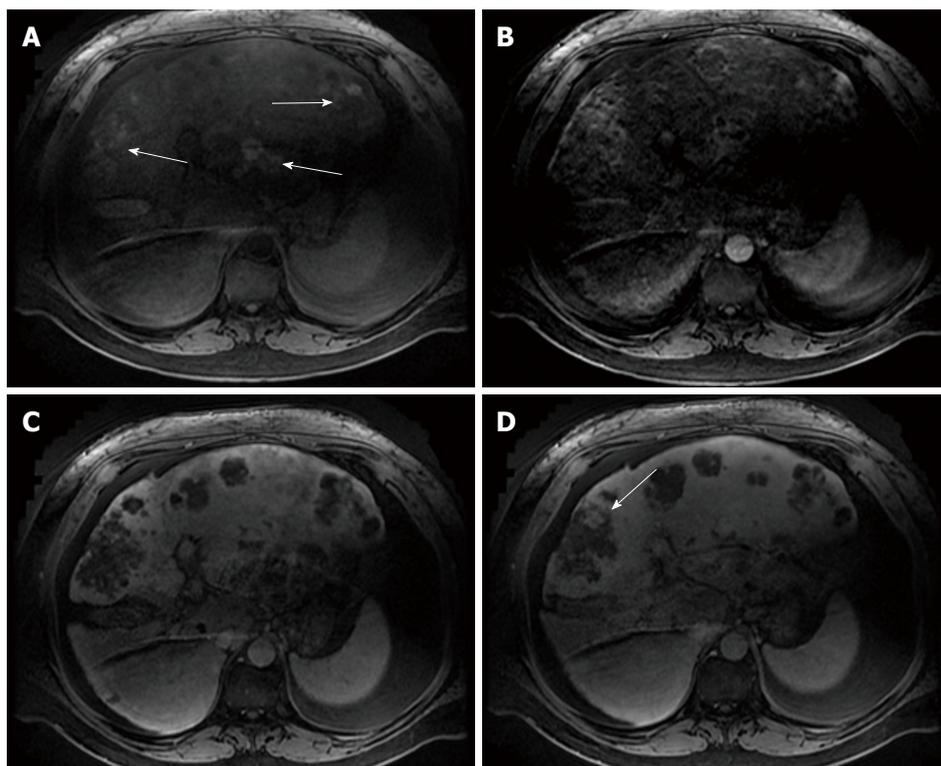


Figure 3 Multiphase axial magnetic resonance images of hepatic angiosarcoma. A: Precontrast image shows multiple mildly intense diffuse masses and areas of increased density in center lesions (thick arrows); B: Arterial phase demonstrates multifocal masses with peripheral enhancement; C: Portal venous phase illustrates multiple, discrete low intensity tumors; D: Delayed phase shows mild persistent enhancement of low intensity tumors (arrow) without definite washout.

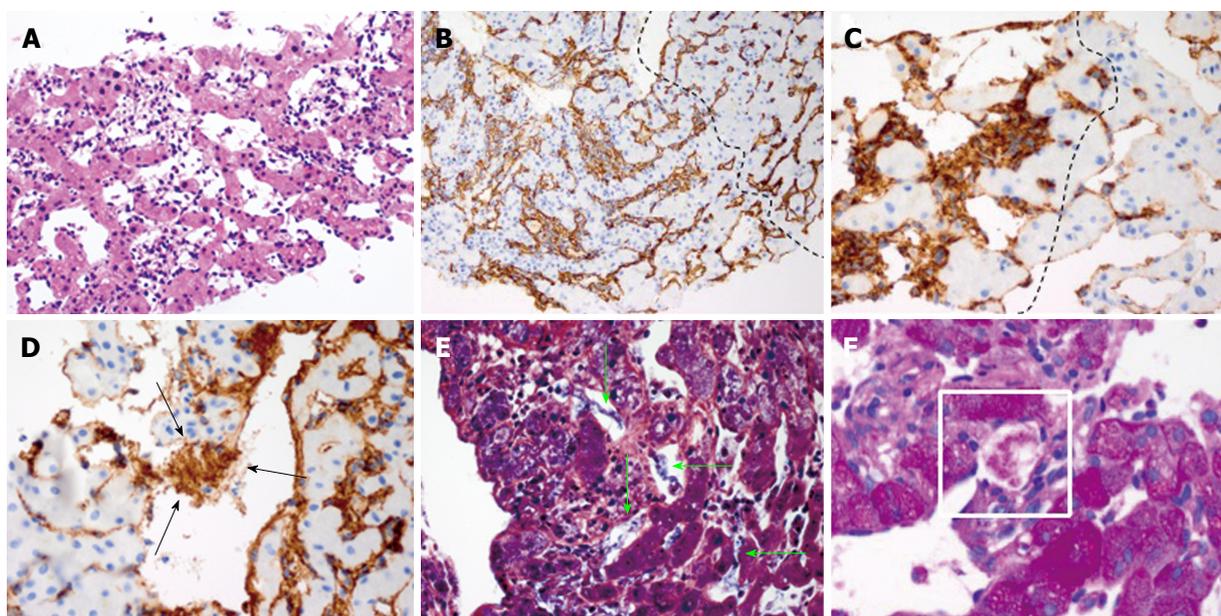


Figure 4 Histopathological findings of tumor needle biopsy suggest the presence of Kasabach-Merritt syndrome. A: Hematoxylin-eosin (H&E) staining of tumor demonstrates dilated vascular channels lined by atypical endothelial cells with hyperchromatic, enlarged nuclei and reticular cytoplasm; B: Immunohistochemical stain for vascular antigen CD34 showing diffuse infiltration of CD34+ cells throughout the sinusoids in the tumor (left of line) with focal aggregation. Uninvolved region (right of line) shows normal liver sinusoidal endothelial cells (LSEC) that highlight the nondilated sinusoids along the cord of hepatocytes; C and D: Immunohistochemical stain of von-Willebrand Factor (vWF)/Factor VIII shows increased expression within tumor cells (left of line) as compared to uninvolved region (right of line). Extracellular aggregate positive for vWF/Factor VIII is seen within dilated sinusoid of angiosarcoma (arrow); E: Phosphotungstic acid-hematoxylin stain (PTAH) demonstrates fibrin nets within the tumor seen as extracellular fibrillary structures that stain blue (arrow); F: Periodic acid-Schiff (PAS) stain of the tumor demonstrates glycogen granules within extracellular material of vascular channels, representing clumps of entrapped platelets (shown in rectangle). Note that positive PAS staining of glycogen is also observed in native hepatocytes.

angiosarcoma (Figure 4C-F). Of importance, there was no histological evidence of cirrhosis. These findings strongly suggest hyper-activation of the coagulation cascade as well as entrapment of platelets within the tumor. Therefore, our histological findings are congruent with the proposed concept of KMS.

There are 72 case reports of KMS to date. Of those, 43 cases were associated with hemangioma, 16 with Kaposi hemangioendothelioma/tufted angioma, 8 with angiosarcoma, 2 with lymphangioma, 2 with angiolipoma, and 1 with Merkel cell carcinoma. These cases demonstrated marked abnormalities in coagulation and thrombocytopenia. However, none of these reports provided histological validation of KMS and therefore it remains indistinguishable from tumor-associated DIC.

Our histological investigation proposed the potential mechanism of hyper-activation of the coagulation cascade *via* up-regulation of vWF/Factor VIII within the dilated sinusoid of the tumor. Moreover, we speculate that the upregulated vWF/Factor VIII results in the downstream formation of fibrin nets and subsequent entrapment of platelets within the tumor. Our findings highly suggest that this is the potential explanation for the significant thrombocytopenia and deregulated coagulation cascade. Thus, our report provides a conceptual advancement for the differentiation of tumor-associated DIC from the systemic manifestation of coagulopathy occurring within the vascular tumor. In conclusion, we report a case of idiopathic hepatic angiosarcoma with features of KMS with clinical and histological evidences.

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COMMENTS

Case characteristics

A 40-year-old man with no past medical history presented with a 2 mo history of worsening abdominal pain and jaundice.

Clinical diagnosis

Distended abdomen with diffuse tenderness and scleral icterus without signs of chronic liver disease.

Differential diagnosis

Hepatocellular carcinoma, metastatic adenocarcinoma, cirrhosis, disseminated intravascular coagulation (DIC).

Laboratory diagnosis

Anemia, thrombocytopenia, decreased albumin, elevated bilirubin, elevated

INR, decreased fibrinogen, elevated d-dimer.

Imaging diagnosis

Multiphase computerized tomography and magnetic resonance imaging showed discrete, multifocal, and isodense masses involving all segments of liver, largest measuring 6.5 cm.

Pathological diagnosis

Angiosarcoma with tumor cells strongly expressing CD34 with uninvolved region showing no evidence of chronic liver disease.

Related reports

Kasabach-Merritt syndrome (KMS) has been reported before to occur in association with some adult vascular tumors. However, none of these reports provides histological validation of KMS and thus the disease entity remained indistinguishable from tumor-associated DIC.

Term explanation

KMS is characterized by thrombocytopenia and hyperconsumption of coagulation factors within a vascular tumor. KMS has been well described in the pediatric population but rare in vascular tumors of the adult population.

Experiences and lessons

This entity can be confused for DIC due to similar abnormalities in coagulopathy. However, by histological investigation KMS can be differentiated from DIC.

Peer-review

It is really the first time to describe a case of primary hepatic angiosarcoma with the clinical and histological evidence of KMS.

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