

Kasabach-Merritt 증후군을 동반한 거대 간 혈관종: Tc-99m 적혈구 간 및 전신 혈액풀 신티그래피와 SPECT소견

전북대학교 의학전문대학원 핵의학교실¹, 내과학교실², 임상의학연구소³
손명희^{1,3} · 정환정^{1,3} · 임석태^{1,3} · 김동욱^{1,3} · 임창열^{2,3}

A Giant Hepatic Hemangioma Complicated by Kasabach-Merritt Syndrome: Findings of Tc-99m RBC Scintigraphy and SPECT Including a Total Body Blood Pool Imaging Study

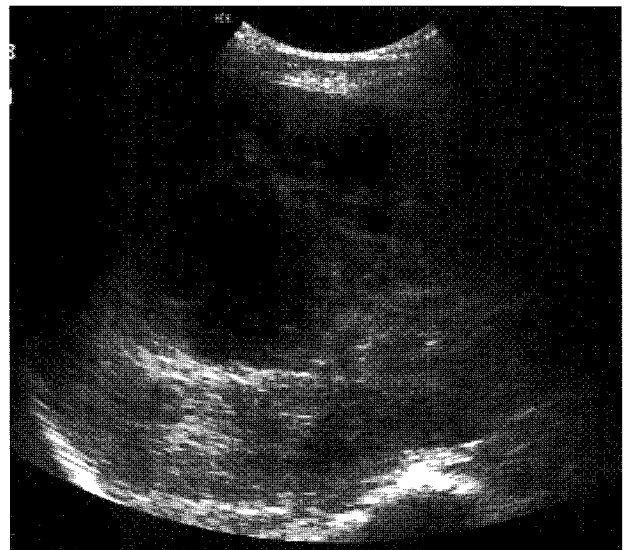
Myung-Hee Sohn, M.D.^{1,3}, Hwan-Jeong Jeong, M.D.^{1,3}, Seok Tae Lim, M.D.^{1,3},
Dong Wook Kim, Ph.D.^{1,3}, and Chang-Yeol Yim, M.D.^{2,3}

Departments of ¹Nuclear and ²Internal Medicine, and the ³Research Institute of Clinical Medicine, Chonbuk National University Medical School, Jeonju, Jeonbuk, Korea

Kasabach-Merritt syndrome (KMS) consists of thrombocytopenia, microangiopathic hemolytic anemia, and localized consumption coagulopathy that develops within vascular hemangioma. This syndrome may also be associated with occult hemangiomas located at various sites. Tc-99m RBC scintigraphy and SPECT have proven to be reliable for confirming or excluding hemangioma. Total body blood pool imaging study during the scintigraphy also provides a means of screening for occult lesions. The authors report the case of a 29-year-old man who presented with a giant hepatic hemangioma complicated by KMS, and underwent Tc-99m RBC scintigraphy and SPECT including a total body blood pool imaging study. (Nucl Med Mol Imaging 2009;43(1):83-86)

Key Words: Tc-99m RBC scintigraphy, Kasabach-Merritt syndrome, giant hemangioma, thrombocytopenia, coagulopathy

Figure 1. A 29-year-old man was admitted because of right upper quadrant pain of 1 month duration, and a painful swelling in the right buttock, which occurred after an intramuscular injection administered 1 week previously. An ultrasonographic scan of the abdomen demonstrated a huge hyperechoic mass with internal hypoechoic areas in the right hepatic lobe. Further evaluation by dynamic MRI was then recommended.



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- **Address for reprints:** Myung-Hee Sohn, M.D., Department of Nuclear Medicine, Chonbuk National University Hospital 634-18, Geumam-dong, Duckjin-gu, Jeonju 561-712, Korea
Tel: 82-63-250-1174, Fax: 82-63-255-1172
E-mail: mhsohn@chonbuk.ac.kr

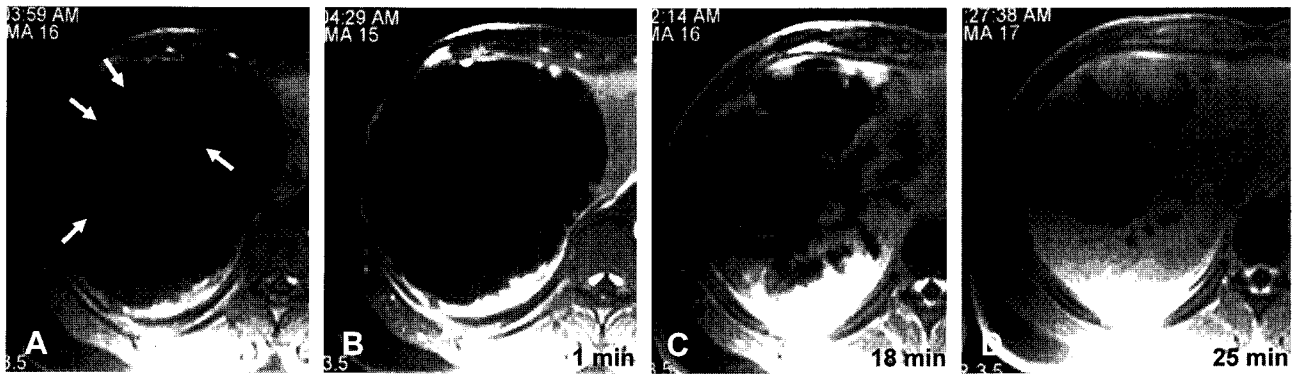


Figure 2. (A) T1-weighted MR image showed a huge, low signal intensity mass in the right hepatic lobe with multiple areas of high signal intensity (arrows) suggesting clot formation within the mass. (B-D) Dynamic contrast-enhanced T1-weighted MR images demonstrated a peripheral enhancement followed by progressive central fill-in of the mass; the typical enhancement pattern of hemangioma. Laboratory findings revealed; WBC 9,800/ μ l, hemoglobin 9.6 g/dl, and platelet 75,000/ μ l. His reticulocyte count was 8%. A peripheral blood smear revealed polychromasia with numerous fragmented red blood cells. His prothrombin time (1.6 INR) and partial thromboplastin time (40.8 sec) were prolonged. His plasma fibrinogen (138 mg/dl) and antithrombin III (25%) levels were low, and total bilirubin (2.2 mg/dl), lactate dehydrogenase (662 IU/L), and fibrin/fibrinogen degradation product (>100 μ g/ml) levels were elevated. The patient was diagnosed as having KMS composed of a giant hepatic hemangioma with consumptive coagulopathy, thrombocytopenia, and microangiopathic hemolytic anemia.^{1,2} This syndrome may also be associated with occult hemangiomas located at various sites, such as, the liver or spleen, head or neck, extremities, and trunk, and may lead to life-threatening bleeding. Tc-99m RBC scintigraphy and SPECT using total body blood pool imaging study were also performed.

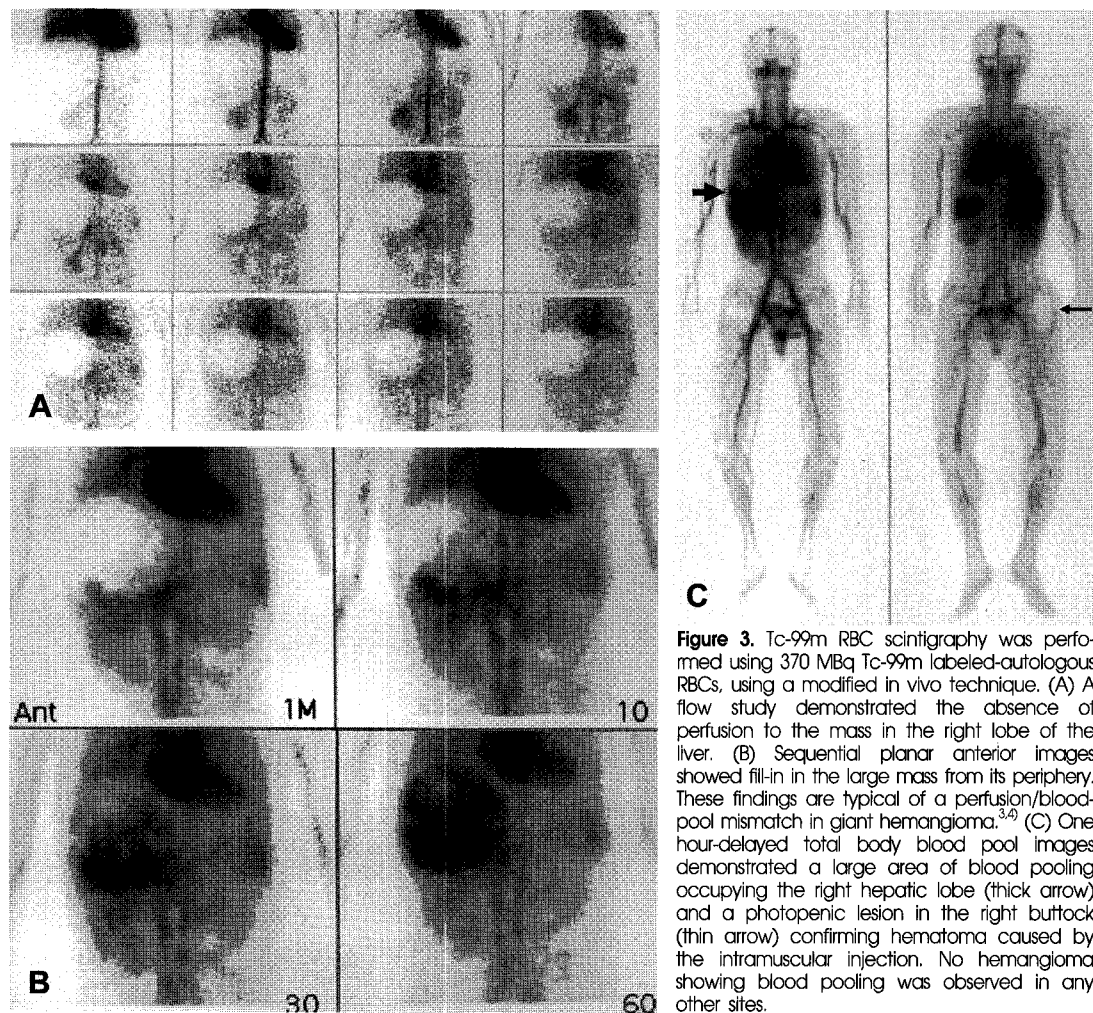


Figure 3. Tc-99m RBC scintigraphy was performed using 370 MBq Tc-99m labeled-autologous RBCs, using a modified in vivo technique. (A) A flow study demonstrated the absence of perfusion to the mass in the right lobe of the liver. (B) Sequential planar anterior images showed fill-in in the large mass from its periphery. These findings are typical of a perfusion/blood-pool mismatch in giant hemangioma.^{3,4} (C) One hour-delayed total body blood pool images demonstrated a large area of blood pooling occupying the right hepatic lobe (thick arrow) and a photopenic lesion in the right buttock (thin arrow) confirming hematoma caused by the intramuscular injection. No hemangioma showing blood pooling was observed in any other sites.

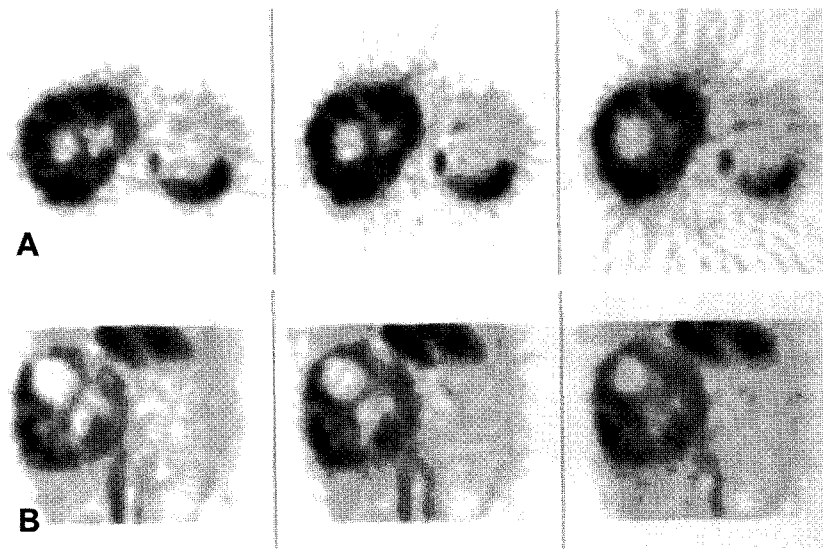


Figure 4. Selected SPECT images on the (A) transaxial and (B) coronal planes obtained after the completion of the late planar image, showing blood pooling in the mass with internal photopenic areas indicating clot formation, as demonstrated by MRI.

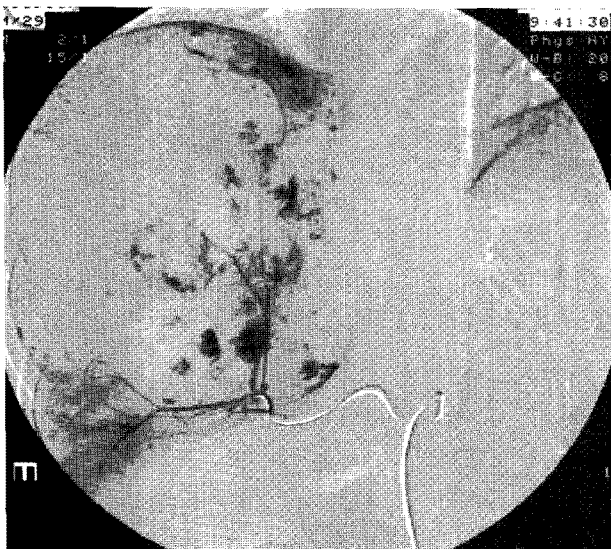


Figure 5. During hepatic angiography, a "cotton-wool" appearance was observed within the mass immediately after contrast injection, which is consistent with a giant hemangioma.⁵⁾ Embolization with Ivalon was performed.⁶⁾ The etiology and pathogenesis of KMS remain largely undetermined. KMS has highest incidence in infants, usually during the first few weeks of life,¹⁾ but it may also appear later.⁷⁾ Various diagnostic procedures have been used to establish the presence of a hemangioma.^{8,9)} However, Tc-99m RBC scintigraphy and SPECT have been proven to be cost effective and reliable diagnostic methods for confirming or excluding the presence of hemangioma.¹⁰⁻¹²⁾ Total body blood pool imaging studies performed during RBC scintigraphy are useful for detecting or excluding occult hemangiomas in other parts of the body.^{13,14)} This report describes a patient with giant hepatic hemangioma complicated with KMS who underwent a total body blood pool imaging study in addition to regular RBC scintigraphy and SPECT.

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