A Huge Subcutaneous Hematoma in an Adult with Kasabach-Merritt Syndrome

Kuan-Lin Wu
Chiung-Ying Liao
Chen-Kuang Chang
Shang-Yun Ho
Yeu-Sheng Tyan
Yuan-Chun Huang

Patient: Male, 33-year-old Taiwanese male.
Final Diagnosis: Kasabach-Merritt syndrome with widespread hemangiomas and an infected huge hematoma in the right thigh.
Symptoms: Gross hematuria.
Medication: —
Clinical Procedure: CT-guided drainage • blood transfusion
Specialty: Hematology

Objective: Rare disease.
Background: Kasabach-Merritt syndrome is a potentially fatal disease that consists of hemangioma(s) with thrombocytopenia, microangiopathic hemolytic anemia, and coagulopathy. Extensive hemangiomatosis is rare. We present the radiological features and treatment strategy of a young adult suffering from Kasabach-Merritt syndrome with widespread hemangiomas and an infected huge hematoma in the right thigh.

Case Report: A 33-year-old Taiwanese male presented with a painful 20-cm mass over his right thigh and gross hematuria for 2 days. Hemangiomatosis was biopically proven in infancy and the patient was under regular follow-up. Physical examination revealed normal heart rate, respiratory rate, and body temperature. Multiple palpable lumps with brown and purple areas of skin over the neck, trunk, and right thigh were noted. Laboratory examinations revealed thrombocytopenia anemia and elevated fibrin degradation products. There were no signs of sepsis. Blood transfusion and steroid therapy were executed. Computed tomography showed a huge complicated subcutaneous hematoma in the right thigh. Drainage of the huge hematoma was performed and antibiotics were prescribed. After the local infection in the right thigh and the bleeding tendency were controlled, the patient was discharged in a stable condition two weeks later.

Conclusions: A huge infected hematoma and widespread hemangiomas are extremely rare complications of Kasabach-Merritt syndrome. There are no known treatment guidelines currently available. Our patient was successfully treated with steroids, drainage, and antibiotics.

MeSH Keywords: Hematoma • Kasabach-Merritt Syndrome • Thrombocytopenia

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/901947

Corresponding Author: Yuan-Chun Huang, e-mail: feberhuang@gmail.com
Conflict of interest: None declared
Background

Hemangioma is a relatively common benign vascular tumor. It is the most common soft tissue tumor in infancy [1]. Kasabach-Merritt syndrome (KMS) is a rare complication of large hemangioma(s) that includes thrombocytopenia and coagulopathy, and can also occur in kaposiform hemangioendothelioma (KHE) and tufted angioma [2–4] (Table 1). We present the radiological features and treatment strategy of a young adult suffering from KMS with a complicating huge hematoma in his right thigh.

Case Report

A 33-year-old Taiwanese male presented with a painful 20-cm mass over his right thigh. Hemangiomatosis had been bi-optically proven in infancy and the patient was under regular follow-up. Multiple palpable lumps with brown and purple areas of skin over the neck, trunk, and right thigh were noted on physical examination. Hematological investigations revealed normal white blood cell count (8,500/mL; N-56%), normocytic anemia (hemoglobin 8.2 g/dL), bleeding tendency with thrombocytopenia (platelet count 80×10⁹/L), hypofibrinogenemia (45 mg/dL), and a prolonged prothrombin time (prothrombin time, 16.9 seconds; control, 12 seconds) with elevated fibrin degradation products (>10×10³ µg/mL). Blood culture results were negative. Chest x-ray showed multiple soft-tissue-density mass lesions at the right axilla, right neck, and left chest wall (Figure 1). Computed tomography (CT) with contrast enhancement showed multiple lobulated masses of varying size involving the right cervical region, right axilla, left chest wall, right paraspinal region, left psoas muscle, peritoneal cavity, and right thigh (Figure 2). These masses showed delayed and progressive centripetal filling in the enhancement pattern, and some masses had phlebolith and cystic components (Figure 3). There was a huge hematoma with septation and peripheral rim enhancement at the right thigh (Figure 4).

A diagnosis of KMS and a complicated huge hematoma in the right thigh was made on the basis of the clinical, radiological, and hematological findings. CT-guided drainage of the complicated hematoma in the right thigh was performed; dark reddish fluid was drained, and a culture was obtained. The culture yielded Staphylococcus lugdunensis, and intravenous antibiotics (piperacillin/tazobactam, 2/0.25 g, every six hours) were prescribed. A transfusion of 2 units of platelets concentrate and 6 units of cryoprecipitate was conducted to replace fibrinogen and correct the coagulopathy. Subsequently, corticosteroids were used for KMS; the patient was treated with intravenous dexamethasone at a dosage of 0.32 mg/kg/day. Three days later, his platelet counts recovered and the steroid therapy was changed to oral prednisolone (2.0 mg/kg/day) with a gradual tapering of the dosage. After the local infection in the right thigh and bleeding tendency were controlled, the patient was discharged in a stable condition two weeks later.

Table 1. Definition of important terms in this article.

| Terms                        | Definition                                                                 |
|------------------------------|---------------------------------------------------------------------------|
| Hemangioma                   | Benign neoplasm characterized by abnormal proliferation of blood vessels  |
| Hemangiomatosis syndrome     | Large or numerous hemangiomas and vascular malformation in the skin and viscera |
| Kasabach-Merritt syndrome    | A condition associated with vascular lesions; manifests as consumptive thrombocytopenia and coagulopathy |

Elsayes et al. [4].
Hemangiomas account for 7% of all benign soft tissue tumors, and frequently occur in infants and children [5]. The tumor can be located in the skin, viscera, subcutaneous tissue, muscle, bone, and even the synovium [6,7]. In our patient, there were multiple, diffuse infiltrations in the chest wall, back, peritoneal cavity, and right gluteal region.

**Discussion**

Hemangiomas account for 7% of all benign soft tissue tumors, and frequently occur in infants and children [5]. The tumor can be located in the skin, viscera, subcutaneous tissue, muscle, bone, and even the synovium [6,7]. In our patient, there were multiple, diffuse infiltrations in the chest wall, back, peritoneal cavity, and right gluteal region.

**Figure 2.** Contrast-enhanced computed tomography demonstrates lobulated masses (arrows) involving the right neck, right axilla, pleural involvement in addition to infiltration of the chest wall, right paraspinal region, left psoas muscle, peritoneal cavity, and right thigh.

**Figure 3.** Contrast-enhanced computed tomography of the abdomen shows delayed and progressive centripetal filling in the enhancement pattern of the lobulated mass (arrows) at the right longissimus thoracis muscle and right spinalis muscle. (A) Early arterial phase; (B) late arterial phase; (C) delayed phase.
On plain film, hemangiomas may present as soft-tissue-density masses, sometimes accompanied by phlebolith formation. Periosteal, cortical, or medullary changes may also be seen in adjacent bony structures depending on its benign or aggressive nature, including cortical erosion, osteopenia, or sclerosis [8]. The lesions have echogenic characteristics, such as blood flow, under ultrasound images which help to differentiate them from other soft-tissue masses [1]. CT scans may reveal soft-tissue masses with phleboliths in pre-contrast studies and variable enhancement in post-contrast studies. Magnetic resonance imaging (MRI) is a better choice for evaluating the interactions of the lesions with adjacent structures and distinguishing the lesions from malignant soft-tissue masses [9]. CT scans may reveal soft-tissue masses with phleboliths in pre-contrast studies and variable enhancement in post-contrast studies. Magnetic resonance imaging (MRI) is a better choice for evaluating the interactions of the lesions with adjacent structures and distinguishing the lesions from malignant soft-tissue masses [9]. The images on MR demarcate ill-defined hyperintense masses, which contain vascular spaces, on T2W images, and intermediate signal intensity on T1W images. Foci of low signal calcification or areas of thrombosis may be seen. It is important to identify slow-flow lesions from high-flow lesions such as arteriovenous malformations or fistulae using dynamic contrast-enhanced MRI [10].

KMS is a rare complication associated with hemangiomas, and has a mortality rate of 20%–30% during the first few weeks of life [11]. The phenomenon also arises from kaposiform hemangioendothelioma (KH), kaposiform lymphatic anomaly, and tufted angioma (TA), and some malignant neoplasms such as angiosarcoma [12]. The radiological presentations of KH and TA are similar to those of other vasoproliferative neoplasms; however, KH tends to be larger in size with a more ill-defined border and an infiltrative growth pattern. KH usually presents with characteristic flow voids owing to numerous feeding and draining vessels.

The overall mortality rate of KMS is 10–37% due to ulceration and recurrent local and systemic sepsis, and is thus an indication for aggressive treatment [13,14]. The syndrome consists of intravascular consumption, clotting, and fibrinolysis within the hemangioma, resulting in degradation and aggregation of platelets and fibrin and elevation of d-dimers, as in our case. Because of the risk of bleeding, KMS may cause subcutaneous hematoma, subgaleal hematoma [15], intracranial subdural hematoma [16], spontaneous spinal epidural hematoma [17], and renal hematoma in the infant [7]. No known treatment guidelines are followed nowadays. Many methods have been recommended, such as the use of steroids, compression, embolization, interferon, laser therapy, sclerotherapy, chemotherapy,
radiation, or surgery [18]. Alfa-2b interferon has been reported to have a good response in about 80% patients [19,20]. Another recently-reported therapeutic option available for infantile hemangiomas is propranolol, a non-selective beta blocker [21]. The goals of treatment are tumor involution and correction of life-threatening coagulopathy.

**Conclusion**

Different types of hemangioma and other hypervascular tumors can cause KMS with manifestations of consumptive thrombocytopenia, intravascular coagulation, and fibrinolysis. The overall mortality rate of KMS is 10–37%; and there are currently no well-developed treatment guidelines. Although a huge hematoma as a complication resulting from KMS is uncommon, it may cause fatal consequences. We report a case of hemangiomatosis with KMS in a young adult and an associated huge complicated hemATOMA. Our report serves to remind physicians of the potential complications that may manifest with KMS.

**Acknowledgements**

The authors thank the Department of Hematology & Oncology for patient care.

**Statement**

The authors declare that they have nothing to disclose.

**References:**

1. Paltiel H, Burrows PE, Kozakewich HP et al: Soft-tissue vascular anomalies: Utility of US for diagnosis. Radiology, 2000; 214: 747–54
2. Croteau SE, Liang MG, Kozakewich HP et al: Kaposiform hemangioendothelioma: Atypical features and risks of Kasabach-Merritt phenomenon in 107 referrals. J Pediatr, 2013; 162: 142–47
3. Malhotra Y, Yang CS, McNamara J, Antaya RJ: Congenital kaposiform hemangioendothelioma with Kasabach-Merritt phenomenon successfully treated with low-dose radiation therapy. Pediatr Dermatol, 2014; 31: 595–98
4. Elsayes KM, Menias CO, Dillman JR et al: Vascular malformation and hemangiomatosis syndromes: Spectrum of imaging manifestations. Am J Roentgenol, 2008; 190: 1291–99
5. Allen PW, Enzinger FM: Hemangiomatosis of skeletal muscle. An analysis of 89 cases. Cancer, 1972; 29: 8–22
6. Hoeger PH, Helmke K, Winkler K: Chronic consumption coagulopathy due to an occult splenic haemangiomatosis: Kasabach-Merritt phenomenon. Eur J Pediatr, 1995; 154: 365–68
7. Guthrie SQ, Rhodes M, Janco R et al: An infant with Kasabach-Merritt syndrome with associated renal hematoma and intussusception. J Pediatr, 2005; 25: 143–45
8. Ly JQ, Sanders TG, Mulloy JP et al: Osseous change adjacent to soft-tissue hemangiomatosis of the extremities: Correlation with lesion size and proximity to bone. Am J Roentgenol, 2003; 180: 1695–700
9. Teo EL, Strouse PJ, Hernandez RJ: MR imaging differentiation of soft-tissue hemangiomatosis from malignant soft-tissue mass. Am J Roentgenol, 2000; 174: 1623–28
10. Vilanova JC, Barcelo J, Smirniotopoulos JG et al: Hemangiomatosis from head to toe: MR imaging with pathologic correlation. Radiographics, 2004; 24: 367–85
11. Arunachalam P, Kumar VR, Swathi D: Kasabach-Merritt syndrome with large cutaneous vascular tumors. J Indian Assoc Pediatr Surg, 2012; 17: 33–36
12. Wen S, Zhang W, Yang Y, Sun J: Angiosarcoma of the scalp and face associated with Kasabach-Merritt syndrome and disseminated intravascular coagulation. Indian J Dermatol Venereol Leprol, 2016; 82: 96–97
13. Aslan A, Meyer Zu Vilsendorf A et al: Adult Kasabach-Merritt syndrome due to hepatic giant hemangioma. Case Rep Gastroenterol, 2009; 3: 306–12
14. Subash A, Senthil GK, Ramamoorthy R et al: Kaposiform hemangioendothelioma with Kasabach-Merritt phenomenon in a neonate of life-threatening nature: A case report. J Assoc Indian Pediatr Surg, 2015; 20: 194–96
15. Stalder MW, Dorafshar AH, Redett RJ: Calcified subgaleal hematoma with secondary cranial deformity in a patient with Kasabach-Merritt phenomenon. J Craniofac Surg, 2011; 22: 208–11
16. Emre U, Gokmen A, Ozen B et al: Spontaneous subdural hematoma associated with Kasabach-Merritt syndrome: A case report. Turk J Haematol, 2012; 29: 291–92
17. Mizuno I, Nakagawa H, Watabe T, Sugimoto I: Spontaneous spinal epidural haematoma in association with Kasabach-Merritt syndrome. J Clin Neurosci, 1999; 6: 518–20
18. Kim S, Kim TU, Lee JW et al: The perihepatic space: Comprehensive anatomy and CT features of pathologic conditions. Radiographics, 2007; 27: 129–43
19. Hatley RM, Sabio H, Howell CG et al: Successful management of an infant with a giant hemangioma of the retroperitoneum and Kasabach-Merritt syndrome with alpha-interferon. J Pediatr Surg, 1993; 28: 1356–57; discussion 1358–59
20. Wysocki M, Mastowska E, Drapińska I et al: Life-saving therapy with alfa-2b interferon in an infant with Kasabach-Merritt syndrome. Pediatr Surg Int, 2016; 32: 623–28
21. Sidbury R: Update on vascular tumors of infancy. Curr Opin Pediatr, 2010; 22(4): 432–37