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

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Manuscript Preparation E
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Conflict of interest: None declared

Patient: Male, 1
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 bioptically 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

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 (KH) 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 bioptically 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 80×10^9/L), hypofibrinogenemia (45 mg/dL), and a prolonged prothrombin time (prothrombin time, 16.9 seconds; control, 12 seconds) with elevated fibrin degradation products ($>10\times10^3$ 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.

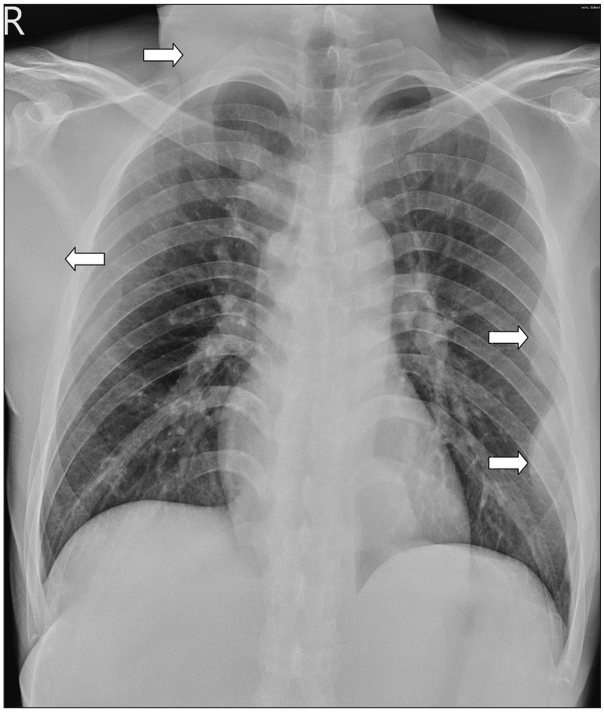

Figure 1. Chest x-ray shows soft-tissue-density mass lesions (arrows) at the right axilla, right neck, and pleural involvement in addition to infiltration of the chest wall.

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].

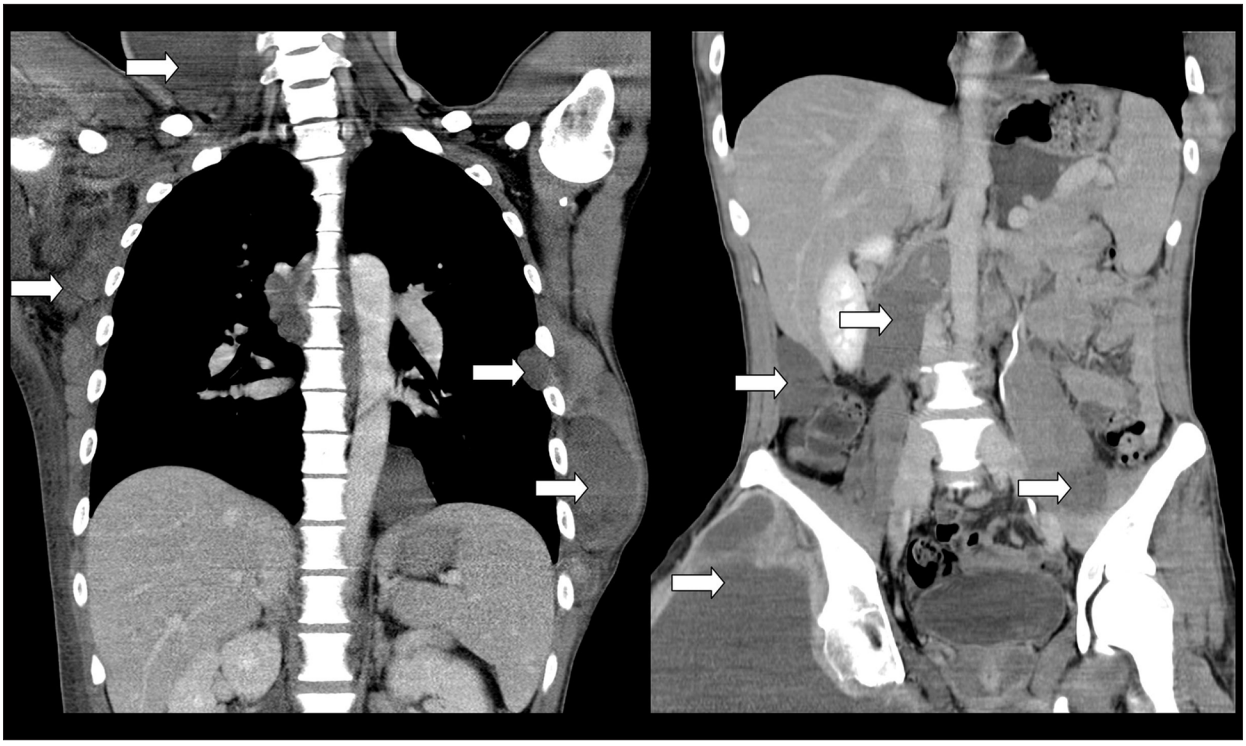

Figure 2. Contrast-enhanced computed tomography demonstrates lobulated masses (arrows) involving the right neck, right axilla, pleural involvement in addition to infiltration of the chest wall, right paraspinal region, left psoas muscle, peritoneal cavity, and right thigh.

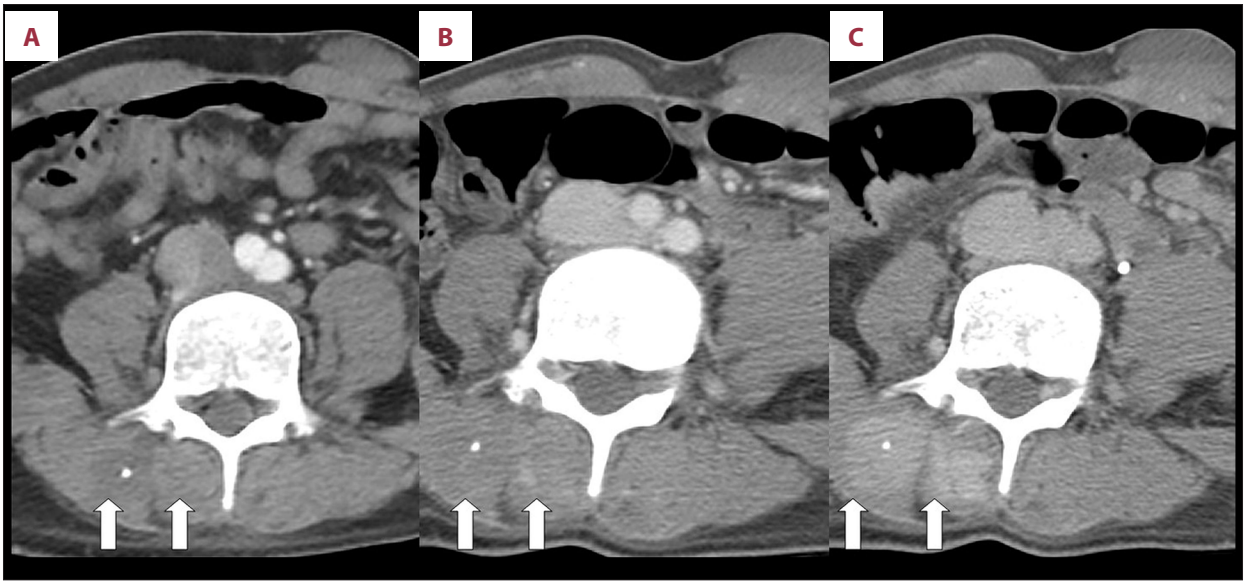

Figure 3. Contrast-enhanced computed tomography of the abdomen shows delayed and progressive centripetal filling in the enhancement pattern of the lobulated mass (arrows) at the right longissimus thoracis muscle and right spinalis muscle. (**A**) Early arterial phase; (**B**) late arterial phase; (**C**) delayed phase.

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.

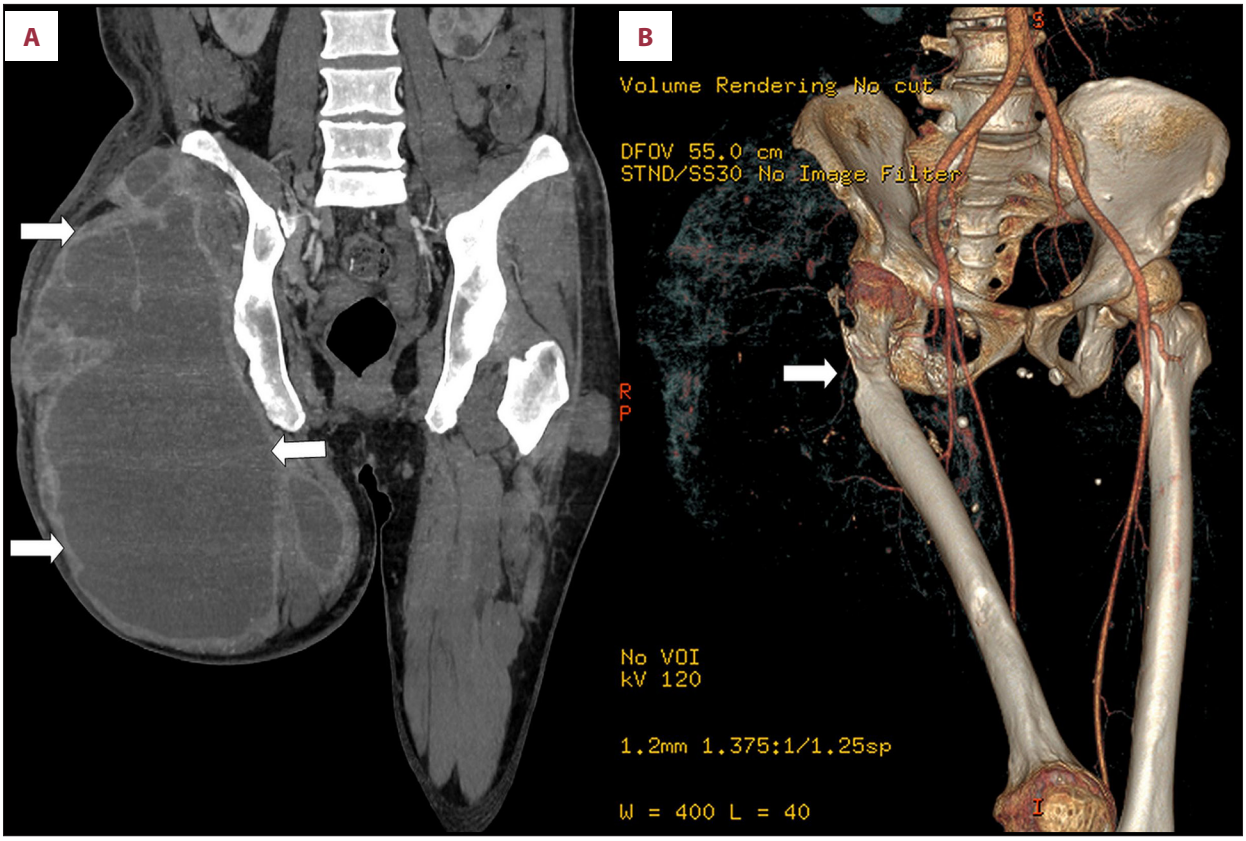

Figure 4. (**A**) Contrast-enhanced computed tomography shows a cystic mass lesion (arrows) with peripheral enhancement and inner septation at right thigh. (**B**) A 3D volume rendering reconstruction shows associated bony erosion of the greater trochanter of the right femur (arrow) due to the mass rather than to osteomyelitis with bony destruction.

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]. 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.

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Statement

The authors declare that they have nothing to disclose.

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