



Unusual cause of acute pulmonary thromboembolism: Kasabach merritt syndrome

Anuradha Rao^{1*}; Chandni Sharma²; Syed Khader Mohammed³; Raghuram P⁴

¹Assistant Professor, Department of Radiodiagnosis, Kidwai Memorial Institute of Oncology, Bangalore, India

²Resident, Department of Radiodiagnosis, Kidwai Memorial Institute of Oncology, Bangalore, India

³Senior Resident, Department of Radiodiagnosis, Kidwai Memorial Institute of Oncology, Bangalore, India

⁴Professor and Head of the department, Department of Radiodiagnosis, Kidwai Memorial Institute of Oncology, Bangalore, India

*Corresponding Author(s): Anuradha Rao

Assistant Professor, Department of Radiodiagnosis,
Kidwai Memorial Institute of Oncology, Bangalore,
India

Email: anu78rao@gmail.com

Abstract

Kasabach-Merritt syndrome [KMS] is a rare but potentially life threatening condition characterized by consumptive coagulopathy caused by hemangiomas. A 64 year old lady was referred to CT scan with history of fever, pain abdomen, respiratory distress, anemia, raised bilirubin levels and altered coagulation profile with clinical diagnosis of disseminated intravascular coagulation. Ultrasound abdomen done in a different hospital showed hypoechoic lesions in liver and spleen and was reported as hepatic and splenic abscesses. CT scan of thorax and abdomen revealed pulmonary thromboembolism with few large hemangiomas in liver and one hemangioma in spleen. Bilateral iliac veins showed thrombus within. A diagnosis of Kasabach Merritt syndrome was made based on imaging and clinical findings. Ultrasound guided fine needle aspiration cytology showed the hepatic lesions to be epitheloid hemangioendothelioma. Thus imaging played a crucial role in the diagnosis of this rare condition presenting as clinical emergency.

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Introduction

In 1940, Kasabach and Merritt reported the association of thrombocytopenic purpura with the presence of a rapidly enlarging capillary haemangioma [1]. Currently the term "Kasabach-Merritt Syndrome (KMS)" has been used to describe coagulopathies associated with enlarging hemangioma, which almost complies with the initial description of the syndrome [1,2]. The mortality rate is said to be between 10 and 37% with more than 80% of cases being diagnosed within the first year of life [3]. A very small group of infants with hemangiomas develop

thrombocytopenia, associated with consumptive coagulopathy. The management plan of these patients mainly focuses on correcting the coagulopathy/ thrombocytopenia and to eliminate the primary cause-the hemangioma. Surgical resection is usually curative [3]. Our case is unique in its presentation as pulmonary embolism has not been commonly reported complication in a patient with hemangioma and DIC. It was the radiologist who suggested the syndrome considering the clinical and imaging findings.



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Case report

A 64-year-old female presented with fever and pain abdomen for 12 days. She had undergone abdominal ultrasound in a peripheral hospital where a diagnosis of hepatic and splenic abscess was made and treated for the same. She was pale with mild icterus. She complained of difficulty in breathing for few hours and was investigated with blood parameters which revealed anemia, raised bilirubin levels, severe metabolic acidosis and altered coagulation profile. The hemoglobin level was 3.5 g/dl, INR was 2.0, prothrombin time was 25 seconds, APTT was 35 seconds, platelet count was 60000/microlitre, and total bilirubin was 2.9 mg/dl. A clinical diagnosis of disseminated intravascular coagulation was made and referred to Radiology department for CT scan of thorax and abdomen. Abdomino-thoracic CT scan revealed pulmonary thromboembolism in the left upper pulmonary artery branches (Figure 1), thrombus in the right brachiocephalic vein extending to the superior venacava (Figure 2), also hypodense thrombus was seen distending the bilateral common femoral veins extending into the external iliac veins (Figure 2). Large hypodense lesions with peripheral enhancement which persisted in venous and delayed phases were seen in almost all the segments of liver (Figure 3); at least 20 in number, largest measuring 84x71mm involving segments 4A, 4B, 2 and 3 of liver. A similar lesion was also seen in the spleen (Figure 3b). Ascites and generalized anasarca was noted. The contrast images were suboptimal probably due to hemodilution with patient's hemoglobin being very low. An imaging diagnosis of multiple hemangiomas in liver and spleen was made with pulmonary thromboembolism, bilateral lower limb deep vein thrombosis. With these imaging and clinical findings a final diagnosis of Kasabach Merritt syndrome was made. Fine needle aspiration cytology showed the hepatic lesions to be epithelioid hemangioendothelioma. The patient was managed conservatively in the intensive care unit for two days in our centre and later once the clinical stability improved was shifted to a tertiary care cardiovascular centre for further management, as our hospital was a tertiary care oncology institute.

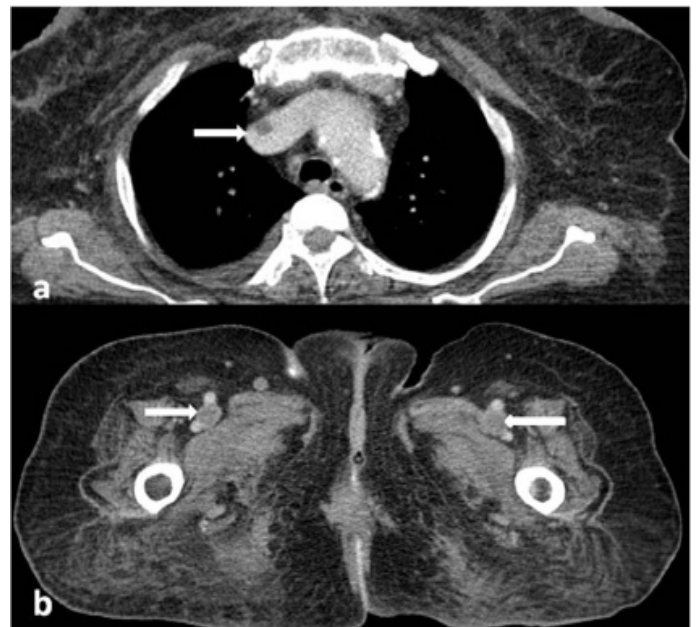


Figure 2: Axial contrast CT of the thorax showing hypodense filling defects [thrombus] in the right brachiocephalic vein extending to the superior venacava [arrow in a]. Also hypodense thrombus was seen distending the bilateral common femoral veins extending into the external iliac veins (arrows in b).

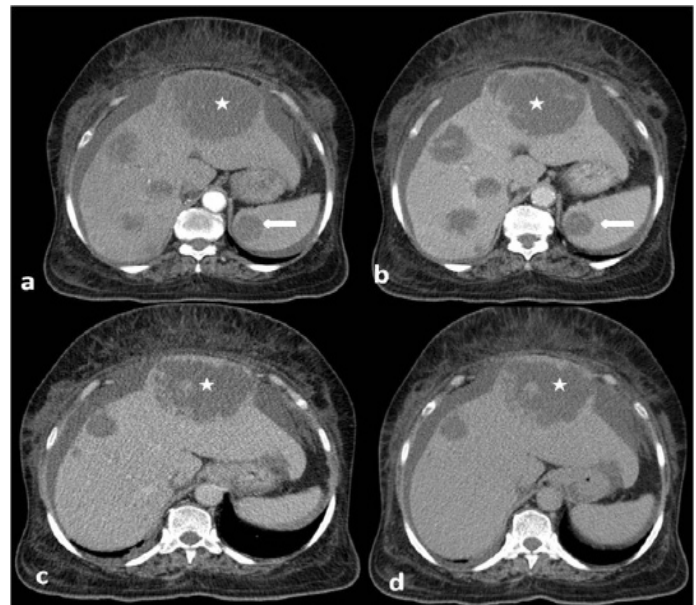


Figure 3: Axial contrast CT of the abdomen showing hypodense multiple liver lesions the largest one showing peripheral enhancement with subtle peripheral filling in of contrast from arterial [a] to portal [b] phases with persistence of contrast in venous [c] and delayed [d] phase. The overall contrast enhancement was not very optimal probably due to extensive hemodilution as the patient's hemoglobin was very low with generalized anasarca. Note also a similar peripherally enhancing lesion seen in the spleen in [a] and [b] (arrows).



Figure 1: Axial contrast CT of the thorax showing hypodense filling defects in the left upper pulmonary arterial branches (arrows).

Discussion

Though the pathogenesis of KMS is not established, the pathophysiology of KMS is attributed to be due to platelet trapping by abnormally proliferating endothelium within the haemangioma resulting in activation of platelets with a secondary consumption of clotting factors. The etiology behind platelet trapping is unclear, and may be attributed to physical entrapment, adhesion to abnormal endothelium within the hemangioma causing aggregation and activation of platelets. Arteriovenous shunting would further increase the level of platelet activation. Thus consumption of platelets and clotting factors causes intralesional bleeding which may be the cause of rapid enlargement of the hemangioma. This becomes a repetitive cycle. International normalized values and D-dimer levels can get elevated. Overall severe altered blood coagulation profile causes Disseminated Intravascular Coagulation (DIC) [4]. Occasional 'spontaneous' resolution of some lesions may be explained by intralesional thrombosis occurring as part of the disseminated intravascular coagulation (DIC)-like picture [1]. There is life long risk of acute episodes of DIC in these patients.

Hemangiomas being the most common benign liver tumor, constitute about 73% of all benign liver tumors and are categorized into 3 types: small (<5 cm), large (5–10 cm), and giant (>10 cm). In patients with giant liver hemangioma, intra-tumoral thrombus can consume a large amount of coagulation factors, which can result in thrombocytopenia and DIC like scenario [5].

Hepatic epithelioid Hemangioendothelioma (HEH) is a rare vascular tumor of endothelial origin with low- to intermediate-grade malignancy with a mortality rate of 20-30% [6]. The HEH is classified as a malignant tumor by World Health Organization. HEH has a prevalence of 1 per 100,000 population. Alpha-fetoprotein, carcinoembryonic antigen and cancer antigen 19-9 are usually normal though liver enzymes can be moderately elevated. Metastases have been reported in 27 %–37 % of patients, the common sites being lung, peritoneum, omentum, mesentery, bone [7]. It can be seen in different patterns- solitary nodular type, multifocal nodular type and diffuse pattern. The peripheral aspect of the lesion, which is rich in tumor cells shows rim enhancement on the arterial phase, while the fibre rich central component can show delayed enhancement. Myxoid, hyaloplasm and fiber compositions are present in the central part of HEH, which determine the CT and MR appearances and also the enhancement patterns. The myxoid and hyaloplasm composition in the central region of HEH demonstrates no enhancement with only peripheral rim enhancement giving the appearance of 'black target sign' or 'bull's eye sign'. This can be confused with metastatic tumors and hepatic abscesses as in our case. When the central part is rich in fiber composition, it demonstrates delayed enhancement and can mimic cholangiocarcinoma. It is difficult to differentiate HEH from hemangiosarcomas and atypical hemangiomas, histopathology being the final mode of diagnosis [8]. Biopsy is complicated due to the high vascular nature of the tumor. Also associated coagulopathy can result in bleeding. Hence before the biopsy, coagulation profile normalization is necessary [9].

Treatments of kasabach merritt syndrome include management of coagulopathy medically before proceeding with surgical or interventional treatments. Corticosteroids, systemic chemotherapy, immunomodulators have been postulated in the treatment. Surgery is recommended in patients with unsuccessful medical treatment [9]. Surgical resection is usually cura-

tive [3]. Liver transplantation is recommended when lesions are extensive [8].

Our case is unique in its clinical presentation where the patient had acute pulmonary thromboembolism. Extensive review of literature has revealed only few cases of pulmonary thromboembolism in a patient with hepatic hemangiomas [10,11]. Most of these cases have reported compression of IVC by the hemangioma as the cause of thrombosis, though a case of recurrent pulmonary thromboembolism by a giant hepatic hemangioma has been reported due to thrombi formation inside the hemangioma [12,13]. Vascular masses associated with Kasabach Merritt syndrome have a disproportionate increased risk of severe bleeding rather than thrombosis, unlike the venous and venolymphatic malformations of the limb like Klippel–Trenaunay syndrome, which have more propensity for thrombosis [14]. Thus the initial acute clinical presentation with pulmonary thromboembolism is rare in Kasabach Merritt syndrome and needs to be considered by the imaging and treating physician alike.

Conclusion

Kasabach-Merritt syndrome is a rare but potentially life threatening condition characterized by consumptive coagulopathy which can be caused by underlying rare vascular tumor like hepatic hemangioendothelioma. Imaging plays an important role in recognition of the underlying cause especially when the initial presentation is acute clinical emergency.

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