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Case Report

Kasabach-Merritt syndrome arising from a vascular fistula

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ABSTRACT

A 58-year-old woman presented with gum bleeding, hematuria, and cutaneous ecchymoses. Left hip replacement had been performed five years prior. The overall findings of our work-up were consistent with ongoing DIC triggered by the presence of an arterio-venous left femoral fistula. The patient was treated successfully with fresh frozen plasma, the fistula was surgically repaired and a stent was placed. On the second day, bleeding had resolved and laboratory values reverted to normal. This uncommon scenario is reminiscent of the Kasabach–Merritt syndrome and well illustrates that patients with an arterio-venous fistula can sometimes present with atypical features. The Kasabach-Merritt syndrome is reported in pediatric and adult patients with giant hemangiomas and angiosarcomas. Adult cases are described also in association with hematomas and large vascular aneurysms. The underlying pathophysiology is the sequestration and consumption of platelets and clotting factors with uncontrolled formation of microthrombi within the vascular lesion. DIC and a microangiopathic hemolytic anemia can subsequently develop. Mechanistic pathways of the Kasabach-Merritt syndrome in the context of a vascular fistula are shared with the more common causes of the syndrome. We speculate that the endothelial dysfunction and injury caused by the flow shear were the pivotal triggers of the aberrant trapping of platelets, the consumptive coagulopathy, and the formation of microthrombi within the fistula. Mortality rate can be as high as up to 40%. The Kasabach-Merritt syndrome could represent the only clinical feature of an otherwise occult vascular fistula. Emergency physicians should be aware of this condition.

To the Editor:

The unexplained onset of a disseminated intravascular coagulation (DIC) is challenging to the clinician. The association of DIC with vascular lesions, i.e. the Kasabach-Merritt syndrome, is poorly understood although its course could be severe and potentially life-threatening [1].

A 58-year-old woman presented with gum bleeding, hematuria, and cutaneous ecchymoses. The patient was taking nimesulide because of hip pain and had hypertension controlled with ramipril and amlodipine. Left hip replacement had been performed five years prior, however surgery was complicated by bleeding and infection and she was taken again to the operating room for revision and hemostasis.

Vital signs were normal. Physical examination revealed cutaneous hematomas, gum bleeding and an enlarged and tense left thigh with no palpably fluctuant mass or crepitus. A thrill was palpated and a vascular murmur was heard. Full blood count showed hemoglobin concentrations 7.8 g/dL, platelets 26 × 10^3 per μL, fibrinogen 70 mg/dL, international normalized ratio (INR) 3.7, and D-dimer 7800 ng/mL. No schistocytes or atypical and dysplastic cells were seen on a blood smear. Angiography showed a left high-flow femoral arterio-venous fistula (Fig. 1).

The overall findings were consistent with ongoing (DIC) triggered by the femoral fistula. The patient was treated successfully with fresh frozen plasma, the fistula was surgically repaired and a stent was placed. On the second day, bleeding had resolved and platelets, fibrinogen and INR values reverted to normal.

This uncommon scenario is reminiscent of the Kasabach–Merritt syndrome [1] and well illustrates that patients with an arterio-venous fistula can sometimes present with atypical features. Diagnosis is delayed and the rates of complications are higher in cases presenting with DIC and no other features suggestive of a vascular fistula such as limb ischemia or high-output heart failure. The Kasabach-Merritt syndrome is reported in pediatric and adult patients with giant hemangiomas and angiosarcomas [2,3]. Adult cases are described also in association with hematomas and large vascular aneurysms [4,5].

The Kasabach-Merritt syndrome is caused by a consumptive coagulopathy resulting in DIC and life-threatening bleeding [6,7]. Mortality rate can be as high as up to 40% [6,7]. The underlying pathophysiology is the sequestration and consumption of platelets and clotting factors with uncontrolled formation of microthrombi within the vascular lesion [6,7]. DIC and a microangiopathic hemolytic anemia can subsequently develop. However, the proportion of patients with DIC and clinically

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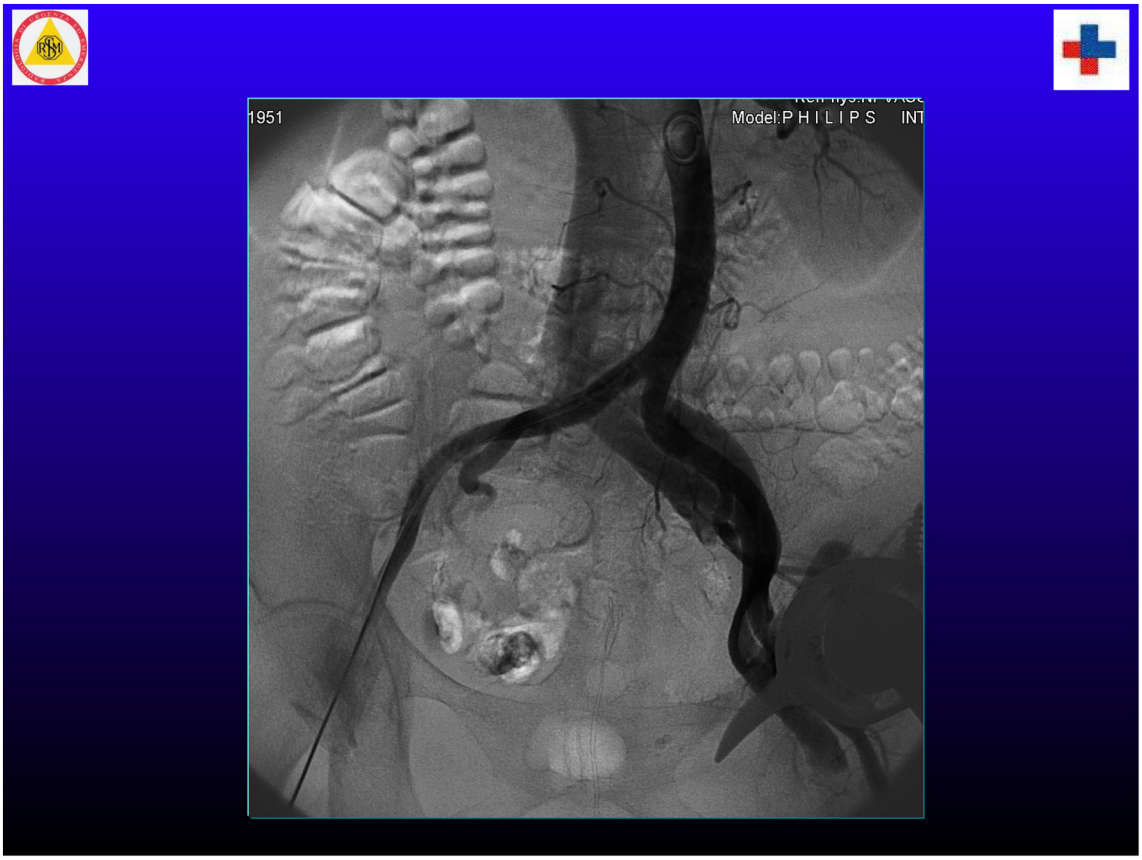

Fig. 1. Angiography shows a high-flow femoral arterio-venous fistula.

relevant bleeding is highly variable with an incidence about 25% among patients with giant hemangiomas and only 0.5 to 1% among patients with large aortic aneurysms [6,7]. There are no figures in patients with an arterio-venous fistula as very few cases have been reported [8].

Our patient does not differ substantially from the classical case series of the Kasabach-Merritt syndrome except for the presence of a small vascular lesion such as a fistula rather than large hemangiomas, angiosarcomas or hematomas. Tying all findings together, we showed a clinically meaningful causal relationship between the vascular fistula and the onset of the coagulopathy. The patient presented with DIC, he quickly recovered after the fistula was repaired, no alternative explanations were found for the onset of DIC, and all other causes of DIC were ruled out upon history, clinical examination, and laboratory and imaging investigation.

Mechanistic pathways of the Kasabach-Merritt syndrome in the context of a vascular fistula are shared with the more common causes of the syndrome. We speculate that the endothelial dysfunction and injury caused by the flow shear were the pivotal triggers of the aberrant trapping of platelets, the consumptive coagulopathy, and the formation of microthrombi within the fistula.

DIC could represent the only clinical feature of an otherwise occult vascular fistula. Our report emphasizes that even small vascular lesions such as an arterio-venous fistula may trigger the onset of a life-threatening consumptive coagulopathy. Emergency physicians should be aware of this condition.

Declarations of interest

None.

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