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A phlegmasia cerulea due to an aortocaval fistula caused by an abdominal aortic aneurysm

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Brief clinical history:

A 58-year-old man consulted at the emergency department for a sudden onset of pain in the right leg. He declared no past medical history except smoking (20 packs.year) stopped 5 years ago. His right lower limb was painful. There was no recent trauma. Vital signs were: afebrile, heart rate of 124 bpm and blood pressure of 156/77 mmHg. The physical examination showed a cold, swelling and blue right leg with marbling (Figure 1). All arterial pulses were found bilaterally. We noted a partial sensitive deficit of the foot up to the knee on the right side and marbling on the abdomen without pain nor bruit.

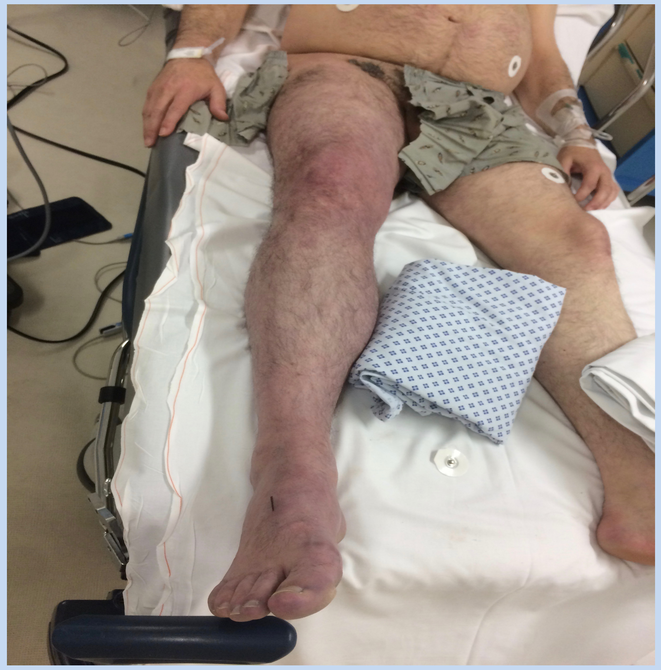

Figure 1: clinical presentation of the cold, swelling and blue right leg with marbling ©NCazes

Misleading elements / Helpful details :

A vascular bedside ultrasound showed a proximal deep venous thrombosis (to the primitive iliac vein up to the popliteal vein) allowing to evoke the diagnosis of phlegmasia cerulea (Figure 2). We tried to see if the thrombosis concerned the inferior vena cava and discovered fortuitously an abdominal aortic aneurysm (AAA).

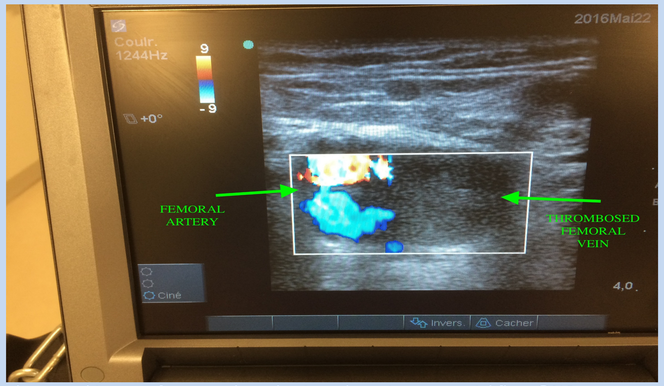

Figure 2 : ultrasound image of venous thrombosis and persistent arterial flow ©NCazes

An enhanced thoraco-abdomino-pelvic CT scan was immediately performed and confirmed a sacciform infra-renal aortic aneurysm of 7 centimeters diameter, partially thrombosed with an aortocaval fistula (ACF) and a bilateral distal pulmonary embolism (Figure 3).

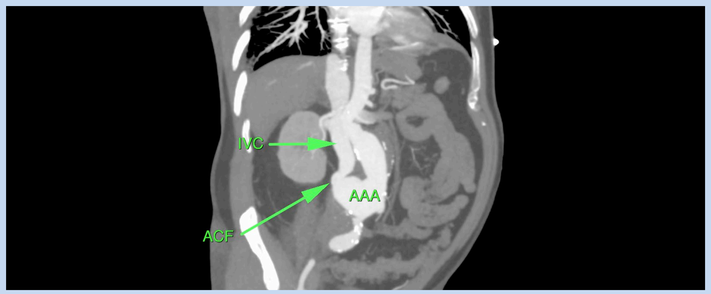

Figure 3 : CT-scan image of aortocaval fistula (oblique coronal view) ©NCazes

Differential and actual diagnosis :

The patient had an aorto-bi-iliac bypass to repair the aorta and a patch on the inferior venacava to close the fistula and has finally totally recovered. AAA disease is a common pathology nowadays in the Western population with a prevalence estimated at 2% to 5% among men aged over 50 years and its spontaneous rupture is correlated with a high mortality. The prevalence of ACF in patients with AAA is low, about 2% to 6% but it remains a well-known complication. The typical described clinical presentation is in fact not so common (<50%) and it is more likely to show the association of an abdominal pain, a high-output congestive cardiac failure and a large venous inflow with its complications (swelling and cyanotic lower limb, hematuria, acute renal failure, scrotal edema, priapism...).

Educational and/or clinical relevance :

In this case report, phlegmasia cerulea was the only symptom of ACF due to AAA. This association of conditions is rare with only one similar case found in the literature. The phlegmasia cerulea is an acute and massive deep vein thrombosis which needs an urgent medical, and sometimes surgical, treatment to avoid necrosis and limb amputation. AAA was compressing the inferior vena cava caused obstruction of venous outflow which was responsible gradually to a diffuse pre-thrombotic state of the lower limb venous network. The total and sudden thrombosis of the common iliac and femoral and veins after the migration of a venous embolism is the classic evolution. The phlegmasia cerulea due to ACF is a rare complication of the AAA but it belongs to these unusual features which emergency physicians should know.