IMAGING IN INTENSIVE CARE MEDICINE

Blue nose syndrome in acquired thrombotic thrombocytopenic purpura

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A 65-year-old man was admitted to the intensive care unit for stupor, tachycardia and gastrointestinal bleeding. Clinical evaluation reported a painful blue nose surrounded by purple infiltration of the perinasal skin (Fig. 1). Other findings included diffuse non-necrotic skin purpura, lack of liver or spleen enlargement, oliguria and atrial fibrillation, as well as anemia (8.2 g per deciliter), thrombocytopenia ($33,000/\text{mm}^3$), mechanical intravascular hemolysis (undetectable haptoglobin, serum lactate dehydrogenase levels superior to ten-fold the normal rate, numerous schizocytes on the blood smear, negative Coombs test) and acute kidney injury (creatinine level 4.04 mg per deciliter, $356 \mu\text{mol/L}$). Fibrinogen and PT were normal. Subsequent laboratory tests were positive for positive antinuclear antibodies (1:400), negative anti-dsDNA and antiphospholipid antibodies, with complement consumption [C3 0.58 (0.8–1.60) and normal C4]. Troponin was increased at greater than 100-fold the normal rate and cardiac echography disclosed segmental changes in contractility of the left heart ventricle and decrease in cardiac output and cardiac contractility. Blood and urine culture were negative. Diagnostic workup for this thrombotic microangiopathy syndrome reported undetectable ADAMTS13 activity and positive IgG anti-ADAMTS13, therefore confirming the diagnosis of acquired thrombotic thrombocytopenic purpura. Skin biopsy documented thrombosis of microvessels with intracapillary thrombi, but no arteriolar inflammation and no skin deposits of C3 or IgG (Fig. 1).

Fig. 1 Blue nose (profile panel *a*, front panel *b*) surrounded by purple infiltration of the perinasal skin. Panel *c* discloses the skin biopsy showing thrombosis of microvessels with intracapillary thrombi, but no arteriolar inflammation and no skin deposits of C3 or IgG

Plasmapheresis and steroids were started immediately after ICU admission for this case of severe thrombotic thrombocytopenic purpura with cardiac, neurological, and renal involvement. Such a cutaneous appearance, especially involving the nose, has never been described in patients with thrombotic microangiopathy.

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