Schnitzler syndrome mimicking septic shock: a challenging presentation for an easy treatment

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BACKGROUND

Schnitzler syndrome is a rare underdiagnosed acquired auto inflammatory disease associated with monoclonal IgM component presenting with neutrophilic urticarial dermatosis, recurrent febrile rash, leucocytosis, severe anemia, and bone/joint involvement. The pathophysiology is still unclear, but relationship with an altered cytokine pattern and mutations of NLPR3 inflammosome has been hypotized. The IL-1 receptor antagonist ANAKINRA seems the only therapy that gives a rapid and long lasting improvement of sintomatology.

CLINICAL CASE

A 74-year old man with an anamnesis of Schnitzler syndrome, was admitted to our hospital due to an acute ischemic cerebral stroke. During the admission he received one unit of red blood cell for anaemia (7g/dL) and after two hours he suddenly developed fever (40°C) with shaken chills, hypotension (70/45 mmHg), tachycardia (128 bpm),desaturation (90% in room air), oligo-anuria, mottled skin and lactic acidosis. We ruled out an adverse reaction due to blood transfusion starting with fluid resuscitation with cristalloid (30ml/kg) and performing triptase, urine an blood samples cultures. Before the administration of the IV broad spectrum antimicrobial therapy, a complete normalization of the clinical picture occurred with optimal gas exchange (pH 7.4 PaO2/FiO2 >250), normal blood pressure (115/75), body temperature (36.5 °) and urine output, reduction of blood lactates and resolution of the mottled skin. The coltures resulted negative with PCT 0.2 ng/ml. The patient was discharge from ICU the day after reporting other episodes of sudden auto-resolving fever never linked to the blood transfusion that he received previously for intercurrent anaemia.

CONCLUSION

Acute and severe inflammatory event may occur in Schnitzler syndrome mimicking a septic shock onset. However, the prompt auto-resolution and the negative cultures ruled out any infection source.

References : **Schnitzler Syndrome**: a Review. Gusdorf L, Lipsker D. Curr Rheumatol Rep. 2017 Aug;19(8):46.