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Case Report

About a Case, Macrophagic Activation Syndrome Post Vaccination

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Abstract: *Introduction:* Macrophage activation syndrome (SAM) results from inappropriate activation of macrophages in the bone marrow and lymphoid organs, responsible for hemaphagocytosis. It is a clinicopathological entity characterized by an excessive and uncontrolled inflammatory response that can be life-threatening. We report the case of macrophage activation syndrome complicating pleuropulmonary tuberculosis. *Observation:* this is the case of a 36-year-old patient, with antimalarial vaccination against malaria for a trip to the endemic area, who presents himself 4 days after his vaccination, in a table of asthenia and NYHA stage III dyspnea. the clinical examination found a conscious patient GCS 15/15, generally impaired, tachycardia at 125bpm, hypotensive at 85 / 32 mmHg, tachypneic at 36 cpm, with SpO2 at 87% in the open air, biologically: normal normocytic anemia at 8.4 g / dl, thrombocytopenia at 26000 / ul, leukocytosis at 34350 / ul, hyponatraemia at 123 mmol / l, hyperkalaemia at 6.26 mmol / l, uremia: 2.1g / l, cratininemia: 54 mg / l, Hepatic cytolysis with ASAT 3 times normal and ALAT 4 times normal, management consisted in admission to intensive care unit evolution was pejorative within the 24 hours of admission, by the installation of a state of refractory septic shock, The post-mortem myelogram confirmed macrophage activation. *Discussion:* The incidence of SAM is poorly known and its frequency is underestimated. A rare disease, the diagnosis of which is often delayed, especially in rapidly developing patients. The diagnosis remains an emergency for the care that remains non-consensual. *Conclusion:* SAM is a rare and poorly described clinical situation but unfortunately a source of wandering and delayed diagnosis. Rare, severe and often unrecognized can compromise life expectancy. The aim of the treatment is to control the macrophage reaction.

Keywords: Macrophage Activation Syndrome, Hemophagocytosis, Vaccination Antimalarial

1. Introduction

Macrophage activation syndrome (SAM) or hemaphagocytosis syndrome is a clinicopathological entity characterized by an excessive and uncontrolled inflammatory response, often accompanied by a life-threatening multi organ failure. Hemaphagocytosis corresponds to the phagocytosis of figurative elements of the blood: erythrocytes, leukocytes, platelets, their precursors as well as cell fragments, by the cells of the monohistiocytic lineage, hence the Anglo-Saxon term "hemophagocytosis syndrome" [1]. The exact pathophysiology of secondary SAM is still poorly understood, but is associated with hyperproduction of interferon and interleukin-18, responsible for an exacerbated immune response and cytotoxicity defect. Infiltration of tissues by activated cells and inflammation may lead to multivisceral failure [2]. Although reactive SAM may occur in apparently immunocompetent subjects, they are much more frequent in constitutional, acquired or

iatrogenic immune deficiency. It is therefore necessary to look for such a deficit. Nevertheless, regardless of immune deficiency, macrophage activation syndrome is usually initiated by an intercurrent disorder. The most common disorders associated with SAM are infections and lymphomas [3].

This syndrome is particular by its immunological pathophysiological complexity; Its lack of knowledge leads too often to a late treatment that is harmful to the patient because it is linked to a large release of many cytokines responsible for a severe clinical condition often fatal [4]. The clinicobiologic picture is not very specific, and may range from a fever associated with thrombocytopenia to a true multivisceral failure syndrome. This lack of specificity is the cause of many late diagnostic, sometimes in patients whose clinical and biological condition degrades to the point of requiring management in intensive care [5]. Clinically, no signs are specific, it may associate fever, hepatosplenomegaly, adenopathies, cutaneous and neurological manifestations, or pulmonary infiltrates. Biologically, bi- or tricytopenia, hepatic cytolysis, elevation of LDH, and coagulopathy associated with hemaphagocytosis image on a cytological or histological sample. This during association with hyperferritinemia and hypertriglyceridemia is very strongly suggestive of SAM.

We report the case of macrophage activation syndrome complicating anti-malaria vaccination

2. Observation

This is the case of a 36-year-old patient, with antimalarial vaccination against malaria for a trip to the endemic area, who presents himself 4 days after his vaccination, in a table of asthenia and NYHA stage III dyspnea. the clinical examination found a conscious patient GCS 15/15, generally impaired, tachycardia at 125bpm, hypotensive at 85 / 32 mmHg, concentrated urine, tachypneic at 36 cpm, with SpO2 at 87% in the open air and 93% under oxygen glasses, biologically: arterial gas: pH: 7.04, PaO2: 83 mmHg, PCo2: 32 mmHg, HCO3-: 17 mmol / l, SaO2: 90%, lactate at 3.4 mmol / l normal normocytic anemia at 8.4 g / dl, thrombocytopenia at 26000 / ul, leukocytosis at 34350 / ul, thick drop: negative hyponatraemia at 123 mmol / l, hyperkalaemia at 6.26 mmol / l, hypocalcemia at 76 mg / l, uremia: 2.1g / l, cratininemia: 54 mg / l, Hepatic cytolysis with ASAT 3 times normal and ALAT 4 times normal, Viral, HIV and CMV hepatitis were negative, on chest x-ray: imainterstitial syndrome, management consisted in admission to intensive care unit where the patient benefited from non-invasively ventilation, on the haemodynamic level: 2 Fluid bolus at 20cc / kg of normal saline 0.9% then introduction of noradrenaline to have a MAP at 65mmhg and corticosteroids, evolution was pejorative within the 24 hours of admission, by the installation of a state of refractory distributive shock, The post-mortem myelogram confirmed macrophage activation.

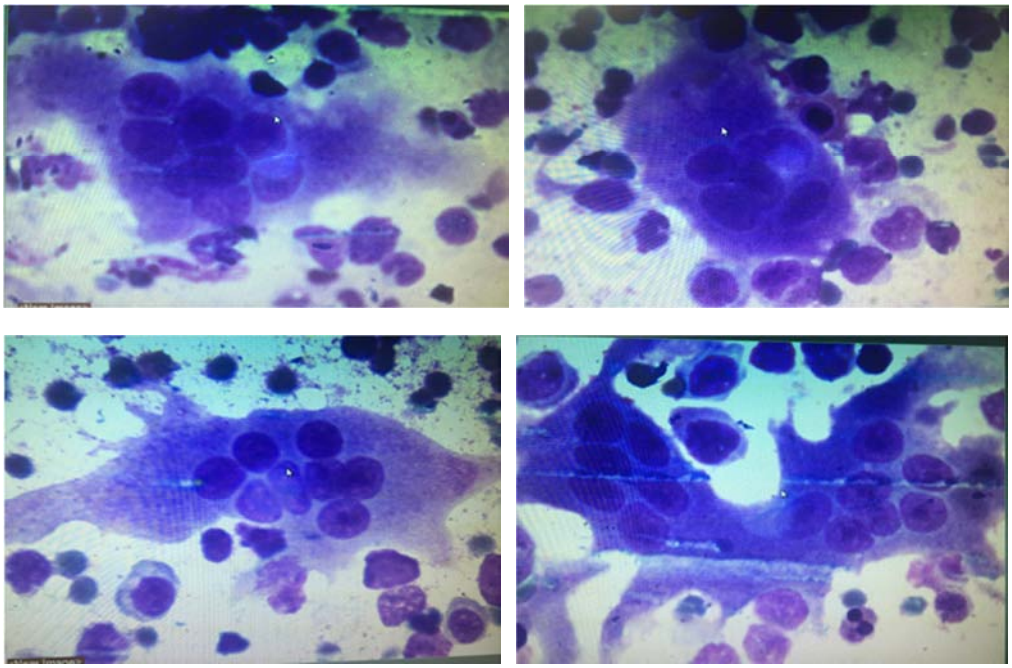

Figure 1. *Myelogram with May-Grümwald-Giemsa staining (1000 × magnification). Image of hemophagocytosis: activated phagocytic macrophage of figured elements of the blood (lymphocytes, erythroblasts, platelet, metamyelocytes and red blood cells) surrounded by a clear halo.*

3. Discussion

The incidence of SAM is poorly known and its frequency is underestimated [5]. The clinical picture is often quite brutal with a sometimes misleading polyvisceral involvement, involving fever, hepatosplenomegaly, peripheral lymphadenopathy, cutaneous eruption, digestive signs or neurological signs, and signs of multivisceral failure. Biological abnormalities are numerous, often major, but not specific. It is their association that leads to evoke the

diagnosis of macrophage activation syndrome [6, 7].

Diagnostic criteria are fever, cytopenia, hypertriglyceridemia and hemaphagocytosis to cytology, with increased transaminases, hyponatremia and hyperferritinemia. This almost completely corresponds to our case or only the dosage of triglyceridemia and ferritinemia were missing [8-10].

SAM is a serious, life-threatening condition whose etiological diagnosis is still not easy [3, 11]. The severity of the prognosis of SAM imposes an aggressive diagnostic approach and a multidisciplinary therapeutic management associating resuscitators, biologists and internists, even before retaining an etiological diagnosis [7, 12, 13].

There are currently no randomized studies in the treatment of SAM. In addition to symptomatic treatment (transfusions, resuscitation) and treatment of the causal disease, the specific therapeutics described in the literature are aimed at decreasing the activation of T lymphocytes and macrophages [14, 15].

4. Conclusion

SAM is a serious, life-threatening condition. It must be evoked in front of any syndrome febrile with cytopenias. Cytology, or even histology, confirms the diagnosis by showing histiocytic infiltration with images of hemophagocytosis. These images may be missing and it is then the clinic that takes precedence and recalls the diagnosis. it remains a severe syndrome, with a dark prognosis, which must be detected and treated without delay, sometimes before the etiological diagnosis. This observation illustrates the diagnostic difficulties of the SAM, and leaves the impression of an uncontrollable acutisation of the SAM, which rapidly evolves towards the multivisceral faillure and to death. Recent advances in the understanding of its cellular origin may make it possible to better adapt treatments and improve the survival of these patients.

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