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Macrophage Activation Syndrome Associated with a Sepsis at the Urinal Starting Point to Escherichia Coli

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***Abstract*—** *Macrophage activation syndrome (MAS) is a rare but potentially fatal disease, it can be primary or hereditary, diagnosed mainly in newborns and infants with a family history, as it can also be secondary, usually found in adults without any notion of family damage, following infections, malignant homeopathies as well as autoimmune diseases. It is an inappropriate proliferation and activation of macrophages in the variable-origin medulla in response to cytokine effects. The diagnosis is based mainly on clinical, biological, non-specific, and cytological or histological criteria, requiring the search for images of hemophagocytosis.We report our first observation of MAS in a nine-month-old infant, secondary to an E coli septicemia, urinary starting point, without any notion of family injury, the clinical picture is dominated by a state of septic shock, hepatosplenomegaly, and general state alteration. This work aims to emphasize the severity of the prognosis and evolution of this pathology, and the difficulties of its management, and to draw attention to any similar or suspicious picture to be prompt and effective in the diagnosis and etiological treatment.*

I. INTRODUCTION

Macrophage activation syndrome (MAS) or hemophagocytic syndrome is a complex clinical-biological association resulting from the inadequate activation and proliferation of macrophages that play a central role in immune defense. It is still unknown and its diagnosis is often delayed in some patients due to the atypical and polymorphic presentation of its manifestations. MAS is a rare but potentially serious disease, its diagnosis involves a cytological or histological study in order to look for images of hemophagocytosis. The so-called "primary" or hereditary MASs that mainly affect newborns and infants with a family history. The “Secondary” MASs, for which no notion of family harm is found, affecting older children or adults. They occur during neoplastic, autoimmune or infectious conditions. (1)

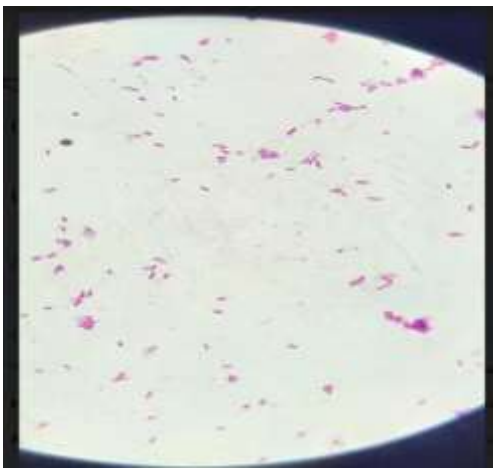
In this work, we reported an observation of MAS to draw attention to clinical, non-specific biological and cytological diagnosis, as well as to highlight its dark evolution, and the difficulties of treatment.

II. OBSERVATION

This is a nine-month-old baby without any family history, hospitalized in pediatric in an intensive care unit, for anemia and infectious syndrome, temperature at 39°C, his general condition is altered with hepatosplenomegaly. On the biological level, the haemogram shows pancytopenia with normocytic normochromic anemia, reticulocytes at 61 G/l, Hb at 5.9 gm/dL, leukocytes at 5200/mm3 with neutropenia at 0.8G/l and thrombocytes at 85000/mm3, as well as the haemogram has been checked on smear.

Hemostatic balance was at normal rates, Fibrinogen was at the lower limit: 1.97g/l. CRP at 98g/l, high ferritin at 601ug/L, triglycerides at 2,2g/ L, liver balance (AST, ALT) at slightly increased rates, renal balance without abnormalities, also for the hemolysis balance, vitamin dosage B12, B9, the self-immunity balance without peculiarities, as well as for the serological balance that was negative. At the ECBU, a germ of Escherichia Coli sensitive to 10 at power 5 was isolated, but the same germ was identified on two hemoculture (aerobic) vials. (pictures1, 2) The myelogram carried out in the laboratory of our regional hospital in Errachidia, shows a cytological aspect of many pictures of hemophagocytosis. (Picture 3). In this way, the diagnosis of MAS associated with a sepsis at the starting point of urine was based on all the above-mentioned biological, cytological, clinical and epidemiological factors.

Picture 1: E. Coli isolated on medium CLED



Picture 2: Gram stain on CLED (BGN : E. Coli)

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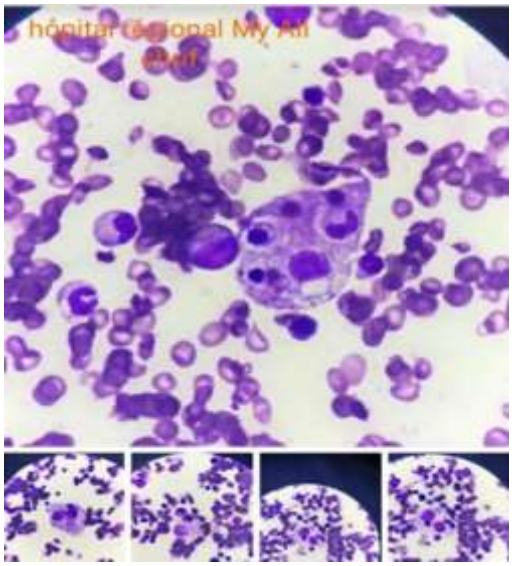
Therapeutically, the infant was given third-generation cephalosporin (10 days) with a bolus of methylprednisolone (3 days), and two transfusions of the armpit and then relayed by oral corticosteroid therapy.

Despite treatment and resuscitation, progress has not been favorable, with a rapid deterioration of his clinical condition that has evolved into multivisceral failure and subsequent death.



Picture 3: Exapmle of hemaphagocytosis on the myelogram

Myélogramme	
Richesse	+, Moelle à densité diminuée
Mégacaryocytes	Rares
Lignée granulocytaire à 57% faite de :	PNN :33% métamyélocytes :14% Myélocytes :8%, Promyélocytes :2%
Lignée érythroblastique :	22%
Lymphocytes :	13%
Monocytes :	4%
Éosinophiles	3%
Plasmocytes	1%
Conclusion :	L'aspiration a ramené un échantillon moyennement riche en cellules, avec rareté des mégacaryocytes, présence de quelques macrophages avec de nombreuses images d'hémaphagocytose, aspect cytologique médullaire d'un SAM.

Picture 4: Patient myelogram

III. Discussions

Macrophage Activation Syndrome is a disease whose prevalence is probably underestimated (1). Its incidence in Japan in 1994 was estimated at 51.7 cases per year, including pediatric and adult cases.

(1). Stephan's study, a systematic sternal puncture in 20 non-immunosuppressed patients, hospitalized in the resuscitation service for a septic shock and having unexplained thrombocytopenia, revealed 60% of cases (2). Laroche conducted a study in France, with the participation of 39 centers. During this study the number of adult MASs for the year 2000, all etiologies confused, was 85 cases, of which 55 of infectious etiology (1). Pediatric forms are often better documented and a Swedish series notes an incidence of one case per million children per year (1). MAS can occur at any age. All populations are affected, but the frequency of associated conditions may vary depending on the population (2). The sex ratio is variable depending on the authors; there is a female predominance for Albert and Coll and a male predomination for Reiner and Coll (2). The diagnosis of MAS is based on the combination of clinical, biological, non-specific and histological or cytological signs. It must be systematically evoked in the face of an unexplained multivisceral failure (4). MAS is a severe syndrome characterized by the generally rapid onset of an intense fever at 39–40 °C, accompanied by a deterioration in the general state, which responds poorly to antibiotic therapy. The worsening is often rapid, which can lead to the transition to resuscitation, often with a complex, infectious, cancer, hematological history.

Macrophage tissue infiltration will result in the rapid onset of hepatosplenomegaly. Hematological damage appears to be the most common (1). The cardinal sign of this impairment is the presence of cytopenia secondary to intra- medullary phagocytosis of the hematopoietic elements. (5). Anemia was in 83.3% of the lakhman hassan series, and was represented by 86.96%, 82.6%, 81.4%, 100% of the cases in the Tiab, Tsuda, Karras and Diaz studies respectively. (1, 6, 7, 8).

In the literature, thrombocytopenia is often less than 100,000 platelets/mm3 (6). In fact, the appearance of the haemogram in our case was in accordance with these series studies and literature. The signs of hemophagocytosis are very often searched on the myelogram but much less frequently in the nodules or spleen (2). The myelogram is the reference examination; it brings the morphological criteria for the diagnosis of MAS (5). In practice, the most relevant criterion for MAS seems to be the detection of macrophages with images of hemophagocytosis without a percentage notion. The etiological background can sometimes be mentioned on the myelogram when there is a homeopathic, lymphomatic infiltrate (9). Frequently early hypertriglyceridemia, which can reach rates of more than 10 times the normal (1, 7), occurs in 20% of cases (6). In the Laroche study, hypertriglyceridemia was observed in 68.4% of patients versus 93% in Diaz's. In general, it is more than 2g/dL. More characteristically, it is accompanied by an increase in very low-density lipoprotein (VLDL). It corresponds to a lipoprotein lipase deficiency, inhibited by TNF-alpha (5).

It allows to monitor the activity of the disease and normalizes during healing (7, 9). Hyperferritinemia is almost constant, and this increase is sometimes dramatic, multifactorial (inflammatory syndrome, hepatocellular necrosis) (1), and most often exceeding 10 times the norms (6).

IV. Conclusion

MAS is a rare clinical, biological and anatomopathological entity, but with a certain morbidity that can put the vital prognoisis at stake. This condition is characterized by excessive and inappropriate activation of the macrophages causing hemophagocytosis. Primary and secondary MASs (post-infectious, neoplastic, systemic diseases) are distinguished. His diagnosis is an emergency that requires the search for images

of hemophagocytosis and signs of macrophage activation, as well as to install early etiological and symptomatic treatment.

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