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A SINGLE CELL SUMMARIZING THE PATHOGENESIS OF CYTOPENIAS IN HEMOPHAGOCYTIC SYNDROME

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The fulminant hemophagocytic syndrome is an acquired disorder most frequent in children and characterized by fever, jaundice, multiple organ failure, cytopenias, coagulopathy and hypertriglyceridemia. It has been observed in many different clinical settings, such as reaction to drugs, bacterial or viral infection, immunodeficiency, systemic lupus, Still's disease and malignancies.[1,2] A familial form of the hemophagocytic syndrome, known as *familial lymphophagocytic lymphadenopathy* or *familial erythrophagocytic lymphohistiocytosis,* has also been described.[3] The hemophagocytic syndrome is often fatal in both its acquired and congenital form. The etiology of *fulminant hemophagocytic syndrome* is still the object of debate. In contrast, the pathogenesis of cytopenias is well known, since the most striking morphologic feature in the bone marrow is the abundance of macrophages actively phagocytizing all sorts of marrow and blood cells.[4] The macrophage in Figure 1 summarizes the pathogenesis of cytopenia in this disorder, in that it has phagocytized three different types of peripheral blood cells: one erythrocyte, two platelets and one leukocyte. This picture was taken from a bone marrow film of a young woman who developed a typical fulminant hemophagocytic syndrome with severe pancytopenia after a viral infection.

References

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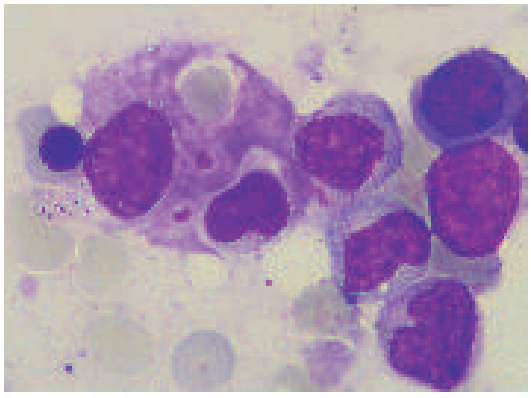

Figure 1. Bone marrow smear, MGG. A single macrophage has phagocytized one red cell, one leukocyte and two platelets.

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