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Hematology

Hemophagocytic Lymphohistiocytosis Revealing a Prostate Adenocarcinoma: About A Case

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Abstract **Case Report**

Hemophagocytic lymphohistiocytosis is a rare and serious condition resulting from inappropriate activation of the immune system. Its association with solid cancers is rare. We report the case of a 58-year-old patient with this rare association and we discuss through this observation the hematological aspects of this condition.

Keywords: Hemophagocytic lymphohistiocytosis - prostate adenocarcinoma - Hematology.

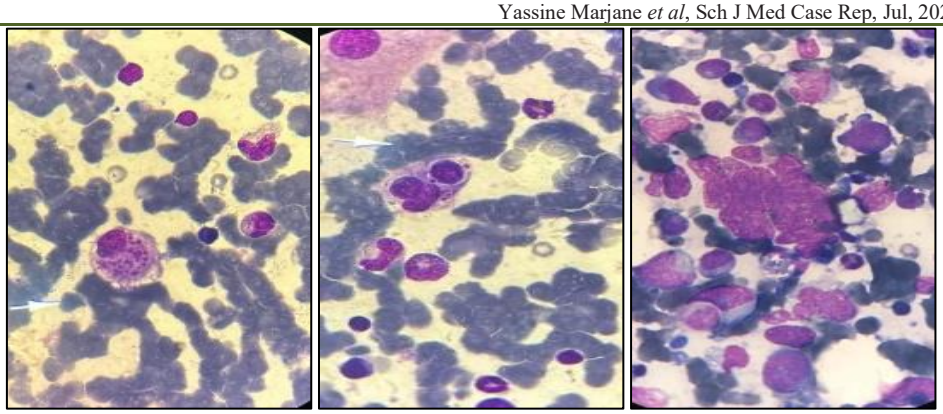
INTRODUCTION

Hemophagocytic lymphohistiocytosis (HLH) is a syndrome of severe hyperinflammation caused by uncontrolled activation of macrophages and T lymphocytes, leading to phagocytosis of the cellular components of blood. HLH is divided into primary forms, related to congenital immune disorders, and secondary forms, triggered by infections, autoimmune diseases, cancers, or neoplastic conditions. Given its high mortality rate, HLH requires prompt diagnosis, although it is complex and relies on a combination of non-specific clinical and biological signs. A bone marrow biopsy is then recommended to confirm findings of hemophagocytosis.

OBSERVATION

A 58-year-old man, with no particular medical history, is admitted to the emergency room for a disturbance of consciousness and acute respiratory distress requiring assisted ventilation. His general condition is altered, with a fever of 39°C, splenomegaly extending beyond the navel, and two left inguinal lymphadenopathies that are hard and painful. Biological tests show bicytopenia with normochromic normocytic arregenerative anemia and thrombocytopenia, hypofibrinogenemia, a decreased prothrombin level, and elevated D-dimers. Biochemistry reveals hepatic cytolysis, an increase in LDH, hyperferritinemia, and hypertriglyceridemia. Serologies are negative for hepatitis B, C, and HIV. Medullary exploration by myelogram reveals a very rich bone marrow with activated macrophages, signs of hemophagocytosis, and clusters of extra-hematopoietic cells.

Tumor markers show an elevation of PSA to 480 ng/ml, and an MRI identifies an infiltrating prostate mass classified as PIRADS 5. The biopsy confirms an acinar prostatic adenocarcinoma of Gleason 9 (5+4), significantly infiltrating the prostate lobes. The patient died two days after the diagnosis.



A B C

Figure: Myelogram obtained after marrow aspiration and spreading, MGG staining (objective x1000) A and B. Hemophagocytosis: macrophage having phagocytosed hematopoietic cells. C. Clusters of extra-hematopoietic cells

DISCUSSION

Hemophagocytic lymphohistiocytosis (HLH) is a state of hyperinflammation related to a "cytokine storm" involving lymphocytes and macrophages. HLH is secondary in 90% of cases, often associated with malignant pathologies, mainly lymphomas, leukemias, and, less frequently, solid tumors with bone marrow metastases [1, 2].

The diagnosis is based on the HScore, which evaluates the risks of secondary HLH by analyzing clinical, biological, and cytological criteria (immunosuppression, fever, organomegaly, certain biological markers, and hemophagocytosis in the bone marrow biopsy) [2].

In the case presented, LHH is associated with metastatic prostate cancer, a correlation rarely described in the literature. The bone marrow aspirate shows no signs of dysmyelopoiesis, but rather reveals extra-hematopoietic cells related to prostate adenocarcinoma, accompanied by elevated PSA [1, 2 et 3].

CONCLUSION

Hemophagocytic lymphohistiocytosis (HLH) is a rare and serious condition resulting from inappropriate activation of the immune system. Its association with solid tumors accounts for only 1% of cases. Its diagnosis is based on clinical and biological signs, as well as images of hemophagocytosis.

Conflict of Interest: The authors declare that they have no conflict of interest.

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