Macrophage-Activation Syndrome and Solid Cancer

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Abstract

Macrophage activation syndrome (MAS) is a rare but severe anatomoclinical entity associated with a high rate of morbidity and mortality, the association of a solid tumor with MAS remains exceptional, reporting the case of a patient followed for squamous cell carcinoma of the nasal cavity with MAS.

Keywords: Macrophage activation syndrome - solid cancer - chemotherapy.

INTRODUCTION

Macrophage activation syndrome (MAS) also known as lympho-histiocytic syndrome or macrophage syndrome is an anatomo-clinical entity resulting from an abnormal activation of the immune system, which is responsible for a cytokine storm, an anarchic proliferation and massive infiltration of organs by benign macrophages leading to a hyperinflammatory state. Acquired MAS is associated to infection, hematological malignancy, systemic disease, and exceptionally to solid cancer.

OBSERVATION

We report the case of a 57-year-old patient followed for squamous cell carcinoma of the maxillary sinus treated initially with surgery then concomitant radio-chemotherapy, who consults 2 years later at the emergency for confusional syndrome with fever since 10 days. The biological results showed pancytopenia (with GB = 1700/μl, HGB = 6.6 g/dl, PLT = 81000/μl) associated with hyponatremia at 111 mmol/l and hypokalemia 2.7 mmol /l.

A realized myelogram objectified the presence of activated macrophages with multiple images of haemophagocytosis with absence of extra hematopoietic cells; a complement of the etiological assessment of MAS (lipid balance, serum protein and imaging) reveals a bone recurrence of squamous cell carcinoma with spinal localization.

DISCUSSION

The diagnosis of MAS is based on the association of clinical, biological and histological or cytological signs. The clinical and biological signs being no specific are often superimposed on those of the causal disease but their association is significant.
The general signs predominate and the main symptom is the fever which is found in a constant way often accompanied by deterioration of the general state especially asthenia and weight loss whose importance is variable.

The MAS and solid cancer association has rarely been reported in the literature between 1979 and 2010, with fewer than 20 isolated published cases carrying a predominance of metastatic malignant melanoma (3 cases) of gastric cancers (3), and germinal mediastinum tumors (3). At least five cases occurred in an additional infectious context (invasive pulmonary aspergillosis, EBV infection, Escherichia coli septicemia, Clostridium perfringens and Klebsiella), and six cases seemed to be only related to the cancer etiology.

Therapeutically, MAS is a therapeutic emergency, which is essentially based on methylprednisolone boluses, a treatment of triggering factors, avoiding the association of NSAIDs.

**CONCLUSION**

With very bad prognosis, MAS treatment is a cancer treatment (chemotherapy and / or radiotherapy), associated with intravenous immunoglobulin or corticosteroid boluses under antibiotic coverage.

**REFERENCES**