



Multiorgan failure after sickle cell vaso occlusive attack: integrated clinical and biological emergency

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Résumé en anglais

We describe the case of a 30-year-old patient, suffering from composite S/beta + sickle cell disease. He was hospitalized following a vaso-occlusive attack with acute bone pains. Despite an analgesic treatment and transfusion of three units of red blood cells, a non-regenerative anemia appeared within 24 hours. One day later an acute chest syndrome with atelectasis of the left lung and desaturation and multi-organ failure occurred and necessitated the patient's intubation and required him to be placed in an artificial coma. A bronchoalveolar lavage was performed, which eliminated pneumonia but proved, after staining with oil red O, many neutral fatty acid microvacuoles in more than 80% of macrophages, suggesting a pulmonary fat embolism. The hypothesis of a bone marrow necrosis causing a pulmonary fat embolism was discussed and confirmed the next day by the characteristic appearance of the bone marrow. A therapeutic protocol associating iteratively bleeding and red blood cells transfusion was administered on the second day with the objective of maintaining haemoglobin S at less than 20% rate. Successive haemoglobin S assay was applied using a high performance liquid chromatography (HPLC) technique with a quick response within one hour after transfusion or bleeding. This protocol resulted in an improvement in the patient's condition, with a gradual normalization of vital signs and extubation twelve days later and discharge without sequelae twenty-five days later. The succession of rare but serious sickle cell complications anaemia which occurred in this patient could be controlled by adapting the laboratory for the clinical emergency.

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