An unusual case of sarcoidosis in an adult patient with sickle cell disease: Management with methotrexate and low dose of steroid

To the editor: Sarcoidosis is a multisystem disease, which pathogenesis is still only partially known. The prevalence of sarcoidosis is largely different depending on race and ethnicity [1]. The association of sarcoidosis and sickle cell disease (SCD) has been reported in few cases [2]. In June 2011, a 52-year-old-man with sickle/β-thalassemia in a care of our service since 2005 presented with a 2 week history of cough and non-specific chest discomfort, without fever or typical SCD-related. He had been treated with hydroxyurea (20 mg/kg/day) for the past 6 years following an acute chest syndrome; during this time he had sporadic episodes of headache with a brain magnetic resonance documenting silent infarction in the left parietal brain area. The patient was clinically evaluated and biochemistry and hematological parameters were performed. Hematocrit was 37%, hemoglobin levels were 11.7 g/dL, reticulocytes were 350,000/μL, lactate dehydrogenase was 708 U/L, and both transaminases were slightly increased (aspartate transaminase, AST 64 U/L; alanine transaminase, ALT: 55 U/L). The pulmonary vital capacity was significantly reduced (<60%). The X-ray chest documented two lower zone pulmonary opacities, while a chest and abdomen computed tomography (CT) scan revealed lower zone pulmonary infiltrates and multiple lymph nodes in the superior mediastinum, two lymph nodes in parathracheal area, few lymph nodes at the celiac trunk and in the inguinal area. The map of enlarged lymph nodes was confirmed by positron emission tomography (PET)-CT. The histology of the removed PET-CT positive inguinal lymph node was consistent with angiolymphoma. Some of the mediastinal lymph nodes were removed with mediastinoscopy. The mediastinal lymph node biopsy showed the presence of partially confluent, non-caseating granulomas with focal associated deposition of fibrous tissue. The lymph node biopsy was consistent with sarcoidosis (fungal and tuberculosis cultures were negative). Hydroxyurea was stopped and erythrocytapheresis was initiated to prevent severe SCD clinical manifestations [3,4]. Although glucocorticoids are the most commonly used therapeutic agent for sarcoidosis, SCD patients incur the risk of developing severe sickle cell related clinical manifestation during treatment with steroids [5]. In sarcoidosis, methotrexate (MTX) has been proposed as a standard second-line agent in patients that cannot use or do not respond to steroids [6]. Limited data on MTX and SCD are present in the literature. In a group of SCD patients with rheumatic disorders treated with MTX, increased vaso-occlusive crisis has been reported [5]. Therefore, the patient was treated with a low dose of prednisone (25 mg/day) associated with MTX (7.5 mg weekly and folic acid 5 mg/week). After 6 months of treatment, the chest CT demonstrated a reduction in size of both lung infiltrates and mediastinal lymph nodes. Neither sickle cell related acute events nor significant changes in transaminases levels were reported in a 10-month follow-up. This is the first reported case of sarcoidosis in SCD patient treated with low dose glucocorticoid and MTX, in which erythrocytapheresis has been used in clinical management of a SCD patient to prevent SCD acute manifestations.

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