Objectives: This case report is presented to emphasize that the deterioration of certain clinical and laboratory findings of an SLE patient should alert the physician for development of MAS.

Methods: We reported a monogenic lupus patient with MAS clinic with acute pancreatitis, who had c1qA gene mutation.

Results: A 9-year-old male was first referred with photosensitivity and erythematous crusted lesions on his extremities and face while he was 2 years old. ANA was positive (1/1000 dilution) and skin biopsy from lesions was compatible with subacute lupus. Steroid, metotrexate and hydroxychloroquine were administered. Patient's male identity and atypical age for lupus was further evaluated with "whole exome sequencing" method. Complement c1qA gene mutation was detected. Monthly fresh frozen plasma and IVIG treatments were added, which able to control the disease to some extent.

He had acute gastroenteritis and fever, leading to activation of skin and oral mucosa lesions with on bilateral parotitis in his last visit (Figure-A). His body temperature was 39.9 °C. USG confirmed the parotitis. Regarding infectious gastroenteritis, IVIG and plasma with broad-spectrum antibiotics were administered. Despite treatment, high fever persisted. He had hypotension, irritability, and hepatosplenomegaly with abdominal tenderness on physical examination. Abdomen CT was compatible with acute pancreatitis (Figure-B), and he had high amylase and lipase levels. Despite IV hydration with intense therapy, developing pancytopenia, hypoalbuminemia, increase in hepatic enzymes, decrease in fibrinogen and ESR with elevated CRP and ferritin levels were alerts for MAS. Bone marrow aspiration demonstrated hemophagocytosis (Figure-C). Pulse high dose IV methyl prednisolone treatment was started. Clinical and laboratory findings, particularly ferritin level, improved in the third day of treatment. Steroids were gradually reduced while cyclosporine was added. He is under remission with low dose steroid, cyclosporine-A and regular plasma infusions per 3 weeks.

Figure. A. Bullous and crusted lesions on face and around oral mucosa. B. Shortening of the pancreas tail in CT. C. Hemophagocytic cells in the bone marrow.

Conclusion: MAS is rarely seen in course of monogenic J-SLE, but may be serious. It is the first juvenile monogenic lupus case with MAS in the English literature, to best of our knowledge. It is important to distinguish this mortal clinical condition from the flare of disease and appropriate treatment should be started as soon as possible.

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CLINICAL AND EPIDEMIOLOGICAL RELEVANCE OF ALLERGY IN SYSTEMIC LUPUS ERYTHEMATOSUS: AN OBSERVATIONAL STUDY

Giuseppe Alvise Ramirez^{1,2}, Andrea Sorce^{1,2}, Francesca Cicero^{2,3}, Mona-Rita Yacoub², Valentina Canti², Giselda Colombo², Enrica Bozzolo², Lorenzo Dagna^{1,2}. ¹Università Vita-Salute San Raffaele, Milan, Italy; ²IRCCS Ospedale San Raffaele, Unit of Immunology, Rheumatology, Allergy and Rare Diseases, Milan, Italy; ³University of Messina, School and Unit of Allergy and Clinical Immunology, Department of Clinical and Experimental Medicine, Messina, Italy

Background: Altered innate and adaptive immune response is a hallmark of the pathogenesis of systemic lupus erythematosus (SLE). Autoimmune manifestations are a major cause of morbidity and damage accrual and constitute the main target of treatment in patients with SLE. Exposure to medications for purposes of immune suppression or treatment of complications might also synergise with an aberrant immune response and facilitate infections as well as hypersensitivity to non-harmful antigens. Epidemiological data about allergy in SLE are scanty.

Objectives: To describe clinical and epidemiological features of allergy and asthma in SLE.

Methods: One-hundred-thirty-five patients with SLE (classified according to the American College of Rheumatology, ACR, 1990 revised criteria) were enrolled. Retrospective data collection included autoimmune manifestations during the course of SLE and history of asthma. In addition, we recorded timing, clinical features, culprit(s) of the first three hypersensitivity reactions (HyR, together with concomitant infections, SLEDAI-2K and prednisone dose at time of each reaction. Damage accrual was quantitated by employing the SLE International Collaborating Clinics/ACR damage index (SDI).

Results: A history of one or more HyR was found in 40.7% of patients. Immediate-type HyR were most significantly represented (83.6% of cases) and four cases were anaphylaxis (lifelong incidence rate of 67.04/105 persons-years). Respiratory allergy (allergic rhinitis or asthma) occurred in 19 patients (34.5%). Twenty-one out of 55 (38.3%) patients with allergy had ≥ 2 manifestations, five (9.1%) had ≥ 3 manifestations. Drugs were the most frequent culprits, accounting for 81.8% of HyR. Early-onset SLE (before the age of 21) associated with a higher risk of becoming allergic during SLE course compared with late-onset SLE (LR=8.01; $p=0.005$; HR=2.28, 95%CI=1.27-4.11). Patients with allergy onset after SLE (52.7% of cases) developed a second HyR more frequently and earlier (LR=11.49; $p=0.001$; HR=5.19, 95%CI=1.87-14.42). SLE with vasculitic manifestations also was a risk factor for developing HyR (LR=4.00; $p=0.045$; HR=2.13, 95%CI=1.00-4.55). Patients with allergy post-lupus had a SLEDAI-2K >4 and a prednisone dose >5 mg/day in 51.7%, 41.7%, 50% of cases and in 37.9%, 50% and 50% of cases at first, second and third HyR respectively. Concomitant infections were detected in 20.7, 25% and 0% of cases respectively. Patients with ≥ 2 HyR had faster damage accrual compared to patients with ≤ 1 manifestations (median SDI/year=0.04, IQR=0.00-0.11 vs 0.00, IQR=0.00-0.06; $p=0.048$).

Conclusion: Patients with SLE have a high prevalence of allergy, which might contribute to its protean phenotype SLE deflects allergy incidence trajectories towards an increased risk. Allergy, in turn, associates with increased SLE activity, medication burden and infectious complications and might affect the risk of long-term damage accrual.

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