Right heart catheterization confirmed PAH with Mean PAP 58mmHg, LVEDP 8mmHg, PCWP 15mmHg

A diagnosis of Mixed Connective Tissue Disease (MCTD) was made, associated with PAH and pericardial effusion.

He was started on Ambrisentan and Tadalafil for PAH. Hydroxychloroquine and Mycophenolate Mofetil were also added in view of the PAH being associated with CTD. The additional pericardial effusion confers a poor prognosis.

Conclusion: Association of Spondyloarthritides and Connective Tissue Disease is rare. There are very few case reports of their chance association, especially MCTD². Our patient had been exposed to Sulphasalazine, Infliximab, Adalimumab and Isoniazid, all with a potential to induce an auto immune CTD. MCTD features have persisted despite drug withdrawal. This case may suggest routinely checking for ANA in SpA patients prior to initiating anti TNF drugs.

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AB1275

HYDROCEPHALUS IN AUTOIMMUNE DISEASES: A SERIES OF CHALLENGING CASES

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Background: Hydrocephalus can be a rare neurological manifestation but lethal complication of various SAIDs, including systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), sarcoidosis, and primary vasculitis. The commonly used medical management programs based on the etiology of hydrocephalus are anti-inflammatory or anti infectious therapies, while surgical management such as ventriculoperitoneal shunts is effective most of the time (Wei Jetal.,2019).

Objectives: To analyze the diagnosis and managment of hydrocephalus associated with autoimmune diseases

Methods: A retrospective case series study was conducted at Rheumatology department.Cairo university.Files were retrieved from the hospital archives by screening records from Jan 2014to Jan2019. Medical records were screened for data regarding the clinical manifestation and outcomeof hydrocephalus associated with autoimmune disease.

Results: Case (1)Male patient 28 yrs old diagnosed a systemic lupus erythematosus The patient was controlled on low dose steroid and azathioprine. In 2019: the patient presented with headache and blurring of vision. Fundus examination showed grade 2 to 3 papilledema. MRI brain showed: Non obstrcutive hydrocephalus. MRA & MRV: normal laboratory investigation was done and showed consumed c3 & c4. ESR: 35mm/hour. Hb: 12.3g/dl. WBC:6.38 cells/cm3. normal liver and kidney function. Neurosurgery consultation was done and recommended urgent ventriculo –peritoneal shunt. follow up fundus showed improvement of papilledema. The patient was discharged on steroids 30mg and mycophenolate mofetyl: 2gram with improvement of his condition.Case (2)Female patient 22 yrs old diagnosed as systemic lupus erythematosus onset 2010. In 2018 she started to develop quadriparesis started in left side followed by Right side associated with attacks of Tonic colonic convulsions. MRI brain showed: Marked communicating hydrocephalus with markedly reduced cerebral hemisphere white matter. MRV & MRA: normal. Her laboratory investigation showed ESR: 49mm/h. HB: 10.3mg/dl, WBC: 5.9 cells/ mm3. Normal platelet count. normal liver & kidney function. Normal c3 & c4. neurosurgery consultation was done. Urgent ventriculoperitoneal shunt was done with improvement of her condition. The patient was discharged on solupred 20mg along with azathioprine.

Conclusion: VPS along with medical treatment with steroids and immunosuppression represent an effective treatment protocol.

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AB1276

NON-INTERFERON THERAPY OF CHRONIC HEPATITIS C IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS AND SECONDARY SJÖGREN SYNDROME - CASE REPORT

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Background: Hepatitis C virus infection is one of the most significant public health problems in the world. To date, the problem of selection of therapy is very relevant for patients with autoimmune diseases and chronic hepatitis C [1]. Patients with systemic lupus erythematosus (SLE) are forced to take different doses of steroids, often in combination with immunosuppressants (mycophenolate mofetil, azathioprine, etc.) even in the case of long-term remission. Such therapy for many years can contribute to the reactivation of hepatitis C.

Objectives: To describe a clinical case of the effectiveness of the combined use of interferon-free therapy and therapy of steroids in a patient with SLE, secondary Sjogren syndrome and chronic hepatitis C.

Methods: 56 years-old woman was admitted to the rheumatology department of Clinical Hospital No. 1 in December 2018 with the debut of SLE: photosensitivity, aphthous stomatitis, arthritis, pleurisy, sicca syndrome, leukopenia, ANA 1: 2560, antibodies to dsDNA 55.95 IU / ml, positive antiSS-A (Ro). At the same time, it became known that she was infected with hepatitis C virus in 1985 presumably. At the time of hospitalization, anti-HCV was positive, the virus genotype was 1b, the activity of the process was low (HCV RNA 6.6x10⁴, AST 33 U/L and ALT 25 U/L). The patient was prescribed corticosteroids (methylprednisolone 16mg/day) and hydroxychloroquine 400mg/day. In January 2019, after gastroenterologist, hematologist and infectious disease specialist advise, it was decided to conduct the patient a specific interferon-free antiviral therapy for chronic hepatitis C (a 24-week course with asunaprevir and daclatasvir), given the potential long-term glucocorticoid therapy, with the prospect of treating the patient with cytotoxic drugs, and the possibility of reactivation of chronic hepatitis C amid of immunosuppressive therapy for SLE and Sjogren syndrome.

Results: Low-disease activity of SLE was achieved in a month, and after 24-week course of antiviral therapy, there was no increase in SLE activity, and positive laboratory and clinical dynamics were noted.

Conclusion: Thus, the use of interferon-free therapy of chronic hepatitis C in patients with systemic lupus erythematosus and secondary Sjogren syndrome shows possible ways to safe treatment of this disease in patients with diffuse connective tissue diseases.

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AB1277

AUTOANTIBODIES IN NLRP3-ASSOCIATED AUTOINFLAMMATORY DISEASE: A CASE REPORT AND LITERATURE REVIEW

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Background: We present the first report of high-titer autoantibodies in *NLRP3*-associated autoinflammatory disease (*NLRP3*-AID). Because systemic autoinflammatory disease (SAID) is characterised by the lack of autoreactive T-cells or autoantibodies, we made a systemic review on the theme of autoantibody in SAID to clarify this phenomenon.

Objectives: We present the first report of high-titer autoantibodies *NLRP3*-AID, and discuss autoantibody in classical SAID.

Methods: We collected the clinical data of the patient with *NLRP3*-AID who had high-titer autoantibodies, and made a systemic review about autoantibody in SAID.

Results: A 38-year-old Chinese Han patient was definitely diagnosed as *NLRP3*-AID because of cold-triggered urticaria-like rash and fever, arthralgia, bilateral sensorineural deafness, chronic meningitis, high inflammatory marker and de novo *NLRP3* T348M variant. Figure 1 shows pedigree of the patient. Meanwhile, she had positive antinuclear antibody (ANA) with a nucleolar pattern of 1:160, positive anti-β2GPI antibody 54-68 AU/ml (normal range < 20 AU/ml) and positive lupus anticoagulant 1.24-1.29 (normal range ≤ 1.2). Literature review found that 13 articles reported autoantibodies in Familial Mediterranean fever (FMF), and there was no autoantibody reported in hyperimmunoglobulinemia D syndrome (HIDS), TNF receptor-associated periodic syndrome (TRAPS) and *NLRP3*-AID. The prevalence of ANA, anti-dsDNA, RF and anti-CCP in patients with FMF was similar to healthy controls.

Fig. 1. Pedigree of the patient. The black arrow indicates the proband. The asterisks indicate the individuals who had clinical manifestations and carry the *NLRP3* T348M variant.

Conclusion: Patients with *NLRP3*-AID can have high-titer ANA and APLs by accident. If patients with high-titer autoantibodies have characteristic manifestations of SAIDs instead of typical features of autoimmune diseases, we should make the final diagnosis through detailed investigation and genetic testing.

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AB1278

A CASE REPORT OF TOCILIZUMAB INDUCE RAPID REMISSION OF ADULT-ONSET STILL'S DISEASE WITH LIFE-THREATENING INTERSTITIAL LUNG DISEASE AND MACROPHAGE ACTIVATION SYNDROME

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Background: Adult-onset still's disease (AOSD) is a systemic inflammatory condition characterized by fevers, polyarthritides, skin rashes and increased leukocyte and neutrophil counts [1]. Sometimes life-threatening complications such as macrophage activation syndrome (MAS), disseminated intravascular coagulopathy (DIC) can be occurred without response to glucocorticoid and disease-modifying antirheumatic drugs (DMARDs).

Objectives: To describe an AOSD patient with life-threatening interstitial lung disease (ILD) and MAS response to tocilizumab.

Methods: This is a case report at our medical practice.

Results: A 43-year-old man was hospitalized for a 2-week history of fever accompanying skin rashes, polyarthritides, sore throat, leukocytosis (the maximum leukocyte count $37.25 \times 10^9/L$ with 89.9% neutrophils), elevated levels of C-reactive protein (190.9mg/L) and ferritin level was very high ($>15000\text{ng/ml}$). Several antibiotics were consequently used, but fever was on going. Chest CT displayed diffuse interstitial lung disease. Infectious diseases and lymphoma were excluded and AOSD with ILD was diagnosed. However, MAS occurred the 20 days later after the pulse of intravenous MP (MP 500mg×3days) following MP 40mg/day, which cannot stop the situation deteriorating. The minimum total white blood cells, hemoglobin and platelets went down to $0.25 \times 10^9/l$, 53g/l and $18 \times 10^9/l$. Finally, tocilizumab rescued this patient from desperation.

Conclusion: Tocilizumab was effective in AOSD with life-threatening ILD and MAS.

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HPR Measuring health (development and measurement properties of PROs, tests, devices)

AB1279-HPR

A DESCRIPTIVE STUDY RELATED TO THE ADHERENCE BEFORE AND AFTER ENROLLING IN A MULTIDISCIPLINARY EDUCATIONAL PROGRAM

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Background: Rheumatoid arthritis (RA) is an inflammatory, chronic disease. It leads to deformity and destruction of joints through the erosion of cartilage and bone. Patients with RA report to suffer symptoms in hands, joints, swelling, loss of motion, muscle weakness among others. (1). Centers of excellence in RA have proposed a multidisciplinary model of care with an initial diagnosis, treatment prescription and follow-up with a rheumatologist, periodic consultations with a physiatrist, psychologist, physiotherapist, occupational therapy, nutrition and a patient focused program (2). With this model of care, the patient is seen as a whole, and the expectation is to achieve the best results in the management of RA. However, if the patient does adhere the model becomes ineffective.

Objectives: The aim of this to report the attendance to a multidisciplinary model of care for patients with RA that attend to a specialized center in Colombia, before and after enrolling in an educational program.

Methods: We performed a descriptive study. Patients enrolled our educational program in July 2019. In our institution patients are followed-up under T2T standards and a multidisciplinary approach, as part of our model of care they have periodic consultations with a rheumatology, physiatrist, psychologist, physiotherapist, occupational therapy and nutrition. We collected sociodemographic data, DAS28, and compare the attendance to each specialty at the beginning and at 6-month follow-up. Descriptive epidemiology was done, we calculated means, and standard deviations for continuous variables and categorical variables were presented as rates. We compared disease activity and adherence at the beginning of the program and after six months of attendance.

Results: We included 229 patients; mean age was 59 years ± 10 ; 93% were female. At the beginning of our program, mean DAS28 was 2.57 ± 1.19 , from all patients 65% were at remission, 11% at low disease activity 19% at moderate disease activity and, 5% at severe disease activity. Regarding adherence to our model, the medical specialty with the highest attendance was rheumatology (30%) followed by, physical therapy (16%) physiatrist consultation (15%) psychology (13%) and, occupational therapy (11%); the specialty with the lowest attendance was nutrition (8%). After six months of attendance to the educational program, we found an increasing number of patients in remission 67%, low disease activity 15%, moderate disease activity 18%, we did not have patients with severe DA28. Regarding the medical specialties, we found a 3% rise in the attendance to the nutrition consultation and psychology consultation. We did not find statistical association between disease activity and adherence to the model.

Conclusion: These results are a clear example of how an educational program is capable of increasing awareness and improving the clinical outcomes and adherence to a multidisciplinary model for approaching RA. As other studies have shown (3), patient education interventions improve adherence to medication and to attendance to health care specialists.

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