that some developed adverse effects which kept them from continuing it. As we believed that hydroxychloroquine should be continued based on the risk-benefit ratio in case of minor adverse events, such as non-severe drug eruption and gastrointestinal intolerance, we decided two systematic approaches for these problems.

For urticaria and erythematous drug rash, we first performed Drug Lymphocyte Stimulation Test (DLST), then offered oral desensitisation if the result is negative. Five patients agreed to undergo oral desensitisation over 5 weeks with crashed hydroxychloroquine tablets in conjunction with once daily levocertirizine 5 mg at night. Desensitisation courses were started on Monday and patients were informed to contact allergologist if skin rash or pruritus emerged. All the five patients successfully went through the graded increment of hydroxychloroquine, and then continued it without the anti-histamine.

For diarrhoea, hangeshashinntou was added. Hangeshashinto, i.e. a Pinellia heart- draining decoction; in Chinese: ban xia xie xin tang), is widely administered in Japan to prevent diarrhoea induced by chemotherapy and immunosuppressive agents such as irinotecan and mycophenolate mofetil. The mechanism is reported to the inhibitory effect of glucuronidase-mediated de-conjugation. Two patients who stopped hydroxychloroquine due to diarrhoea became able to resume it with hangechashintou.

In conclusion, desensitisation of hydroxychloroquine and co-administration of hangeshashintou resulted in better tolerance of the anti-malarial in Japanese patients without serious side effects.

# Poster session 10: Difficult cases

## PS10:182 NMDA-POSITIVE NEUROPSYCHIATRIC LUPUS MANIFESTING AS WEIGHT INCREASE AND HYPOSMIA

[1]C Pamfil, [1,2]S Rednic, [3]I Catana, [4]M Deac Badaranza, [4]D Balint Gib, [1]P Vele, [5]L Bene, [2,4]L Damian. *[1]Rheumatology, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj, Romania; [2]Center for Connective Tissue and Musculoskeletal Diseases, Cluj, Romania; [3]ENT, Sforait Medical Centre, Cluj, Romania; [4]Rheumatology, Emergency Clinical County Hospital, Cluj, Romania; [5]Laboratory Medicine, Emergency Clinical County Hospital, Cluj, Romania*

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**Background** Among the clinical manifestation of systemic lupus erythematosus (SLE), neurolupus (NPSLE) is one of the hardest to identify. While active systemic diseases usually evolve with weight loss, the increase is generally associated with glucocorticoid therapy and immobilisation and rarely with hypothalamic inflammation. NMDA encephalitis, increasingly recognised, may be paraneoplastic or autoimmune.

**Material and methods** We present the case of a 20 year female student, with an SLE with juvenile onset, at 16, mainly with renal features and malar rash. She was in a prolonged remission on azathioprine, hydroxychloroquine, low-dose aspirin and 5 mg prednisone/day. However, after the exam session, she came for a significant weight increment (over 12 kg in 2 weeks), despite allegedly normal eating, accompanied by daytime sleepiness and hyposmia. The BMI, initially 22.5, increased to 27.1, with a 29.3% of body fat by screening impedancemetry. The analyses revealed a high ESR (90 mm/h), an increased AAN titer (1/1280), with high anti-DNA (1055), and an anti-NMDA titer of 933 IU/mL. All other tests, including lupus anticoagulant, anti-cardiolipin, and beta-2 glicoprotein IgA, IgG, anti-Ro, anti-La, and ribosomal P anti-TPO, anti-thyreoglobulin antibodies, FT4 and TSH, were normal. Cerebral MRI was unremarkable, as well as the organ involvement screening, including genital examination and PAP smear. Olfactory function measurement revealed a threshold score of 6 (normal 7–12) and an identification score of 11 (normal 12–15). She received 3 courses of methylprednisolone and cyclophosphamide courses, with slow weight decrease and normalisation after two months and some correction of hyposmia in more than six months.

**Conclusion** Rapid unexplicable weight gain and decreased olfactory perception in lupus may suggest neurolupus or NMDA encephalitis.

## PS10:184 CONSTRICTIVE PERICARDITIS AS THE FIRST PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT AND LITERATURE REVIEW

[1]H Jethwa, [1]M Rana, [2]J Kitt, [1]J Democratis, [1]S Menzies, [1]A Steuer, [1]S Gindea. *[1]Wexham Park Hospital, Slough, UK; [2]South Buckinghamshire Trust, South Buckinghamshire, UK*

10.1136/lupus-2018-abstract.225

**Objective** Although fibrinous and exudative pericarditis is a common feature of Systemic lupus erythematosus (SLE), found in 62% of lupus patients on autopsy, very few cases progress to (effusive) – constrictive pericarditis. We describe the unusual occurrence of constrictive pericarditis (CP) in a patient with Systemic Lupus Erythematosus.

**Methods** This is a chart review- based report of a lupus patient who had constrictive pericarditis as a presenting feature and a systematic literature review of previously published cases. We searched the English medical literature from 1963 to 2016 using PubMed, for terms: 'systemic lupus erythematosus' and 'constrictive pericarditis'.

**Results** A 49 year old African man presented with several weeks of malaise, weight loss, cough, breathlessness, peripheral oedema and hepatomegaly suggestive of right ventricular failure. An echocardiogram demonstrated features of effusive-constrictive pericarditis. The patient was initially treated for suspected tuberculosis; his symptoms progressed in spite of treatment. Further investigations confirmed positive lupus serology (ANA, anti-dsDNA Ab and anti-Sm Ab, low complement levels) and a raised urine protein: creatinine ratio of 177 mg/mmol. A diagnosis of SLE was established and treatment with Hydroxychloroquine and Prednisolone was initiated. However, the CP was refractory to medical management eventually requiring Pericardiectomy.

A literature review identified six other cases of lupus patients with CP. Of these, four patients were male and average age was 38 years. CP was the presenting feature in four cases and TB was part of the differential diagnosis in five cases. The progression from exudative to constrictive pericarditis ranged from one week to six months. Pericardial biopsies performed in four cases showed non-specific chronic inflammation and fibrosis. CP resolved with corticosteroid treatment alone in one case; pericardectomy was necessary in the other five cases for symptom resolution.

**Conclusions** SLE should be included in differential diagnosis of constrictive pericarditis, especially in 'idiopathic' cases and in the context of poor response to tuberculosis treatment. Pericardial biopsy remains a crucial test in excluding an

infectious or malignant aetiology, although the histological findings are typically non-specific for a diagnosis of lupus-related pericarditis. Pericardectomy is likely to be required for definitive treatment.

## PS10:185 NODULAR LOCALISED CUTANEOUS AMYLOIDOSIS IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS

[1,2]M Deac Badarinza, [1,2,3]S Rednic, [1,2,3]C Pamfil, [2,4]C Baican, [5]A vasilache, [1,2,3]I Filipescu, [1]I Muresan, [1]D Balint, [1,3]L Damian. *[1]Rheumatology, Emergency Clinical County Hospital, Cluj-Napoca, Romania; [2]Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania; [3]Rheumatology, Centre for Rare Musculoskeletal Autoimmune and Autoinflammatory Diseases, Cluj-Napoca, Romania; [4]Dermatology, Emergency Clinical County Hospital, Cluj-Napoca, Romania; [5]Hematology, Oncology Institute, Cluj-Napoca, Romania*

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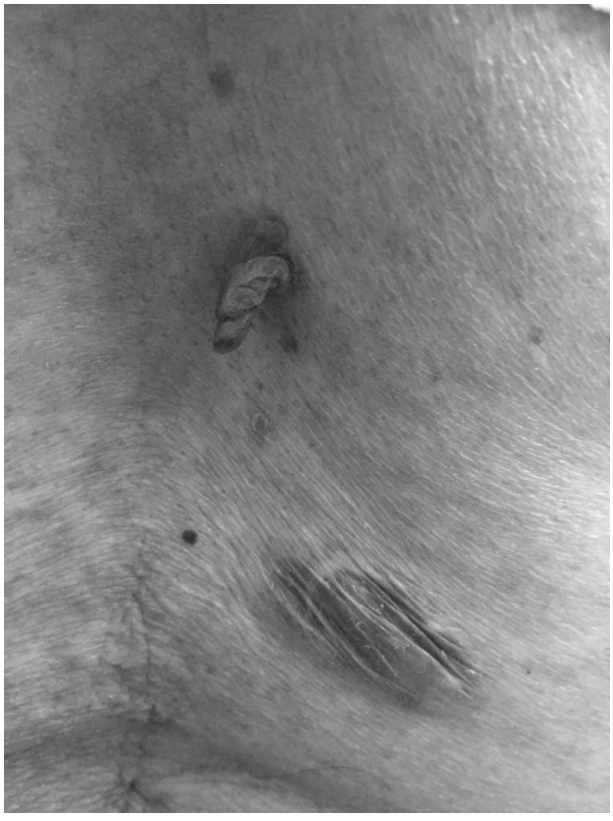
**Introduction** Nodular localised amyloidosis is a rare subtype of cutaneous amyloidosis, associated with various connective tissue diseases, mostly Sjogren's syndrome. Progression to systemic amyloidosis was described in 7%–50% cases. Amyloid deposition was also noted in hypertrofic lupus lesions.
**Purpose** To report a case of systemic lupus erythematosus (SLE) presenting with nodular localised cutaneous amyloidosis, followed up for 17 years.
**Methods** A 55 year patient was addressed to our tertiary unit with pain and swollen in both hands and multiple soft nodular lesions pink to brown on the chest and back.



**Abstract PS10:185 Figure 1**

**Results** Clinical examination and further investigations revealed inflammatory hand arthritis and polyserositis including pericarditis and pleural effusion. Laboratory showed antinuclear antibodies with low anti-dsDNA titer, positive anti-Ro antibodies, positive rheumatoid factor, C3 and C4 consumption. She had negative anti -cyclic citrullinated peptide antibodies and anti-LA antibodies, no sicca symptoms and no ultrasound modification of the salivary glands. The skin histopathology with Congo red staining revealed amyloid deposition in the dermis. A screening for multiple myeloma, including bone marrow biopsy, was negative. She was treated with hydroxychloroquine, and over the time with methotrexate, azathioprine (with loss of tolerance), acitretin (with no significant skin improvement), and topical glucocorticoticoids. New lesions appeared mostly upon cessation of SLE therapy, on traumatised areas and sun exposure, but were quite stable during sustained systemic therapy, suggesting some relation to disease activity. She developed new-onset cryoglobulinemia with increasing anti- Ro titers and rheumatoid factor, but has still normal immunoglobulins, complement fractions and LDH and no light chains on immunelectrophoresis.
**Conclusions** Nodular localised amyloidosis is rare in SLE. The lesions evolve slowly, are minimally influenced by systemic therapy, but a close monitoring for systemic amyloidosis or plasma cell dyscrasia is required even in longstanding cases.

## PS10:187 CAUSES OF DEATH IN CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS IN A TERTIARY CARE CENTRE, SOUTHERN THAILAND

P Vachvanichsanong, E McNeil, P Dissaneewate. *Prince of Songkla University, Hat Yai, Thailand*

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**Introduction** The mortality rate of childhood-onset systemic lupus erythematosus (cSLE) remains high due to the severity of the disease and it complications. The cause of death varies widely depending on the major organs involved and therapy received.
**Objective** To evaluate the causes of death of cSLE.
**Methods** The medical records of children aged <18 years who had been diagnosed with SLE following American Rheumatism Association criteria from 1985 – 2016 in the Division of Nephrology, Department of Paediatrics, Prince of Songkla University, Thailand, were reviewed.
**Results** There were 331 patients, 272 girls and 59 boys, of whom 77 (23.3%) died, 28.6% within the first year after diagnosis. Only 29 medical records were available for evaluation of cause of death. Of these, there were 7 boys and 22 girls with a mean age at presentation of 11.0±3.0 years. The mean follow-up duration was 4.6±3.7 (range 0.2–12.6) years. The major cause of death was sepsis (13) followed by acute respiratory distress syndrome (ARDS) (6), severe heart condition (3), acute kidney injury (AKI) (2), chronic kidney disease (CKD) (2) and intracranial haemorrhage (1). Conditions at the time of death were sepsis (25), pneumonia (16), acute kidney injury (15), bleeding disorders (11), neurological complications (10), ARDS (10), CKD (4), AKI on top of CKD (3).
**Conclusion** The cause of death in cSLE is usually multi-factorial and it is difficult to assign a single dominant cause. Sepsis remains the most common cause of death. In the long-term, end-stage renal disease emerges as an important cause of death in RRT limited institutions.