NODULAR LOCALISED CUTANEOUS AMYLOIDOSIS IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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 hypertrophic 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.

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 glucocorticoids. 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 immunoelectrophoresis.

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.
PS10:189 NEUROPSYCHIATRIC LUPUS. A SEVERE MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

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10.1136/lupus-2018-abstract.228

Purpose Nervous system involvement in systemic lupus erythematosus (SLE) is a grave manifestation of the disease affecting health, quality of life and disease outcome. It is one of the most complex manifestations of SLE and may affect the central, peripheral and autonomous nervous system. Complex interrelated pathogenetic mechanisms are involved in disease pathogenesis. The aim of the study was to describe a patient with neuropsychiatric lupus.

Methods A patient, female aged 50 years presented with SLE with a duration of 20 years. The diagnosis of the disease was made when she presented with intense fatigue, hair loss, a light sensitive rash, arthralgias and positive antinuclear and anti-dsDNA antibodies. In the course of the disease the patient developed CNS involvement with epileptic convulsions, permanent dysarthria and delusions. A brain MRI scan was without specific alterations, however an EEG performed was abnormal and a brain single-photon emission CT (SPECT) revealed decreased perfusion of both frontal and parietal lobes. The patient developed musculoskeletal manifestations at many stages of the disease. At diagnosis the patient had arthralgias of both wrists and knees. The patient presented with a flare with fatigue, mouth ulcers, convulsions, decreased ability to concentrate, intense delusions and dysarthria. At disease flare, when neuropsychiatric symptoms evolved she had diffuse arthralgias.

Results Pulse methylprednisolone i.v. followed by pulse cyclophosphamide i.v. were administered in order to achieve remission. Disease stabilisation was induced by pulse cyclophosphamide at bimonthly intervals and orally administered prednisolone. When remission of the disease was induced by pulse methylprednisolone and cyclophosphamide the patient developed muscle weakness. At disease stabilisation with pulse cyclophosphamide at bimonthly intervals the patient developed arthritis of the hand joints. The disease is now in remission, corticosteroid doses having been significantly reduced.

Conclusions Neuropsychiatric lupus is a grave and complex manifestation of SLE. The disease may be accompanied by various manifestations and severely affects quality of life. Neuropsychiatric lupus should be aggressively treated in order to improve quality of life and disease outcome.

PS10:190 UNUSUAL PRESENTATION OF LUPUS PROFUNDUS

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10.1136/lupus-2018-abstract.229

A middle aged female presented with swelling of face and periorbital region for 4 weeks and fever 3 weeks diagnosed and treated as cellulitis elsewhere examination revealed periorbital puffiness and indurated lesions on face with complete distortion of facial features.

Abstract PS10:190 Figure 1

Swelling of rt masseter, buccinator sterno cleido mastoid
Gross oedema of overlying soft tissue of rt face multiple mildly enlarged ln rt sub mandibular upper and middle jugular erosion of alveolar cortex of rt post edge of maxilla.
Biopsy was done which showed -the subcut. Panniculus shows a lobular inflammatory infiltrate composed mainly of