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

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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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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

Labs-hb- 10.7 gm/dl
Tlc-4500/cumm
Plt- 160×10^3/ microlitre
Sr creatinine-0.8 mg/dl
Lft- sgot –87 u/L, sgpt-64 u/l
Ana if- neg
Ena profile- neg
Apla-neg
La-neg
C3, C4 n
Ct neck with contrast-diffuse enlargement of rt parotid gland
lymphocytes, hyalinising lesions wherein the adipocytes have lost nucleoli are present interspersed, epidermal and dermal histomorphology- follicular plugging present, dermal vessels reveal perivascular lymphohistiocytic infiltrates, stains for afb, pas neg

Opinion- lymphocytic lobular panniculitis with follicular plugging- lupus profundus

Our case was unusual also because of parotid involvement which is a very rare presentation of lupus profundus with very few case reports.

Patient was given steroids and mmf with little response after 6 weeks, mmf was then replaced with thalidomide leading to successful response and resolution of the swelling.