Systemic Lupus Erythematosus

Yet Another Rare Presentation

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Clinical Descriptions

A 14 year old girl was first seen at Karen Grech hospital for complaints of fainting attacks along with occasional convulsions and a progressive depressional state. She was seen by a psychiatrist who thought she had psychomotor retardation and was 'handicapped'. Concurrently, she had complaints of painful and stiff knees, ankles and wrist joints - especially during the morning hours. This went on for sixteen weeks during which period the girl's mental state deteriorated further with her having 'hysterical behaviour' at times, and suicidal tendencies. Her initial resentment of going to school increased and she had finally stopped going altogether for over a month when she was admitted into the Children's Ward with a low grade pyrexia, mental confusion, deteriorating speech and difficulty in walking.

She was a full term baby at birth with a normal delivery in a hospital in Malta. Birth weight 7 lbs. She had no obstetric or neonatal problems and had suffered no significant illnesses in the past. She was never hospitalised before.

On examination at the time of her admission she appeared quite anxious and tense, talking very little and answering only with brief replies. She had a flushed face with a peripheral pulse of 100/min, regular and felt in all the limbs. Blood pressure 140/75 mmHg, oral temperature of 100 F. There was no significant finding in the Cardiac or respiratory system. A detailed neurological examination revealed exaggerated reflexes in all the limbs - more on the right side with down-going planters. Examination of the fundi showed a hypermetropic right disc while the left disc was normal but there was a 'cytoid body' at 11.00 o'clock position. There were no sensory changes and no signs of meningitis or encephalitis.

She also had painful ankles and wrists and had nodular lumps over the dorsum of both her hands. These lumps were present for the last 8 weeks. Hip joint movements were painful on both sides and she had tenderness of the cervical spine.

Detailed investigations revealed a positive Serum Antinuclear factor and L.E. Cells were present in peripheral blood. The R.A. test was negative, serum protein electrophoresis showed a raised level (of 2.9 gm/dl) of gamma globulin, while total proteins and albumin were normal. Serum Creatinine and B.U.N. and urinary excretion of Creatinine during 24-hour period were all normal. A.S.O.T., B.S.R., V.D.R.L. were all negative. E.S.R. was 100 mm in 1st hour (Wintrob). Plasma Cortisol, Serum Prolactin, Serum L.H. and F.S.H. were checked and found normal.

To Summarise

A 14 year old girl presented as case of ? front lobe lesion with history of progressive deterioration of memory, speech and gait. She also had vague joint pains and subcutaneous nodular swellings on both wrists. She had a high E.S.R. of 100 mm in 1st hour, positive L.E. cells and A.N.F. and she responded dramatically to oral steroids. She was rediagnosed as a case of Systemic Lupus Erythematosus.

Discussion

Systemic Lupus Erythematosus is a disease associated with auto-antibodies involving multiple organ systems. The brief list of possible manifestations in the major body systems is given in Table 1.

Table 1

Skin: Butterfly rash, diffuse skin rash, discoid lesions, oral ulcers, alopecia.

Joints: Symmetric tenderness, pain on motion, swelling.

Kidneys: Glomerulitis, Glomerulonephritis.

Serious Membranes: Pleurisy, pericarditis, peritonits.

Blood: Leucopenia, thrombocytopenia, haemolytic anaemia.

C.N.S.: Seizures, psychosis, mononeuritis.

The well remarked *'butterfly rash'* in S.L.E. is frequently absent, but diffuse erythematous skin rashes, discoid L.E. Lesions, or oral ulcers and partial alopecia are frequently seen. The majority of patients with S.L.E. will have one or more of these skin manifestations.

The joint disease is frequently described as *non-deforming* to indicate the difference between a rather mild synovitis and periarticular involvement typical of S.L.E., and the prominent pannus and synovial inflammation of rheumatoid arthritis. Arthritis in S.L.E., is nearly always symmetric and tends to involve essentially the same joints as rheumatoid arthritis. However, visible swelling if often not present, pain is prominent and if any deformity results it is likely to take the form of *Swan-necks* or subluxation, rather than erosive bony destruction.

The predominant features of the renal involvement is in the glomerulus and appears related to immune complex formation, deposition, and the subsequent inflammatory response. In later stages of Lupus nephropatly, arterial involvement, hypertension and uraemia are quite frequent. Renal involvement may be documented by renal biopsy, or by demonstration of proteinuria and appropriate sediment changes together with a low serum complement or antibody to D.N.A.

The commonest haematological finding is the anaemia, which is unfortunately, too non-specific to employ as a diagnostic criterion, but is a useful adjunct in assessment of the progress of the disease. Anaemia most commonly represents a production deficit and reflects the activity of the underlying chronic disease. Coombs positivity is fairly common. Also, some cases present as a thrombocytopenia for which, very rarely, splenectomy may be needed. Leucopenia, with a total white cell count of consistantly around 4000/mm^3 is seen in about one-third of cases.

In the central nervous system, the presenting features may be extremely variable and sometimes confusing. Fortunately, such cases with primarily C.N.S. features are very rare. Seizures and history of fits, sometimes serious emotional disturbances, eg: suicidal tendencies etc., and neuritis have all been documented in S.L.E.

As for the management of these cases, extreme caution should be taken in arriving at the diagnosis especially in patients presenting with complicated multi-system features. Having carefully assessed the patient together with as much of available laboratory reports as indicated, the patient should be commenced on drug treatment. Prior to that a detailed discussion with the patient and the family is essential and the doctor should make a frank attempt to inform them of the exact nature of the illness and its long range implications. In children, and adolescents, bed rest, although very difficult in most cases, will help in the initial phase of illness with painful joints and fever for which Aspirins are useful. Although some authorities advocate using non-steroidal preparations, eg: Indomethacin, the corner-stone of treatment is of course Corticosterodis in moderately severe and severe cases. Steroids are given orally for about 6 to 8 weeks at an approximate dosage of 1mg/kg/day in children - followed by a slow tapering over a 6 month period to half of that level and further reduction as possible. In general, the tendency in recent years has been to taper steroids much more rapidly, and the frankly Cushingoid child is now rarely seen in the out-patients.

Immuno-suppressant drugs, eg: azathioprine or cyclophosphamide treatment remains controversial, and is probably best reserved for specialised units and institutions and the individuals should be thoroughly aware of the inherent hazards of treatment with such drugs.

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