Systemic Lupus Erythematosus – CNS Flare

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ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic, relapsing, remitting, autoimmune disorder having multisystem involvement .The clinical spectrum of SLE is wide and ranges from easily treatable to a very severe life threatening flares involving any organ. Flare can be considered as a reappearance of clinical features, which were earlier, quiescent. Flare involving renal, cardiovascular and central nervous system requires prompt diagnosis and management. We report a case of young female who present as acute psychosis.

CASE REPORT

A 15-year-old girl presented with sudden onset abnormal behavior associated with irrelevant talk and visual hallucinations. She was a diagnosed case of SLE with lupus nephritis for the past one year on prednisolone 10 mg every alternate day, enalepril 5 mg and quarterly doses of cyclophosphamide. There was no history of fever, headache, vomiting, seizures or trauma. However on eliciting history her mother told about her recent result of tenth standard, in which she had failed in four subjects.

On physical examination- the patient had normal vital signs. A cushingoid appearance was noted. Aside from mental status changes – irrelevant talk, irritability, frequent outbursts because of visual hallucinations her neurologic examination was unremarkable.

On laboratory evaluation her CBC was found to be normal. ESR was 55mm after one hour. Urine examination showed no pus cells, no RBCs , albumin was 2+, 24 hour urinary proteinuria -624mgm (same as her previous visit, 2 months back). Blood Urea was 45 mg /dl and Serum Creatine-1.8mg/dl. CT scan of brain showed diffuse cerebral atrophy. However her dsDNA titers were 125 u/ml as compared to the previous value of 45 u/ml.

In light of these findings a diagnosis of SLE with CNS flare was kept. Parental methylprednisolone 1 gm for three days and a repeat pulse of cyclophosphamide 500mg along with oral olanzapine 5 mg once a day were initiated. Within 2 days, a modest improvement in symptoms was evidenced by increasingly appropriate speech and no hallucinations. Patient was eventually discharged to home, completely oriented and appropriately conversant on maintenance oral methylprednisolone, enalapril and olanzapine.

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DISCUSSION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem involvement and diverse manifestations. SLE flare can be considered as a reappearance of clinical features that were earlier quiescent. During flare, organ system may be involved singly or in combination. Flare involving renal, cardiovascular and central nervous system requires prompt diagnosis and management. There are certain triggering factors for lupus flare like, exposure to sunlight, physical and mental stress, intercurrent infections, pregnancy and sudden withdrawal of drugs.

With improved management of lupus nephritis, neuropsychiatric systemic lupus erythematosus (NPSLE), and the complications associated with its therapy have displaced nephritis as a leading cause of mortality in patients with SLE.

Central Nervous System Flare

Headache is the most commonly seen presenting feature, occurring in about 55%, mood disorders occurs in 57%, cognitive dysfunction in 55%, seizure disorder in 51%, acute confusional disorder in 35%, peripheral nervous system impairment in 15%, psychosis in 12%, and stroke in 12%. Psychosis is reported in 8 % of cases and is characterized by delusions or hallucinations.

Lupus psychosis should be distinguished from steroid induced psychosis. Transverse myelitis, demyelination, abnormal movements are rare manifestation seen in only about 1-3 %of cases.

The NPSLE manifestations have been divided into major and minor criteria

MAJOR SYMPTOMS

1) Neurologic

 a) Seizures

b) Focal motor or sensory deficits, stroke, movement disorders, nerve palsies

c) Generalized disturbances: altered consciousness, meningitis, dementia, and possible Organic Brain Syndrome

2) Psychiatric

a) Psychosis, depression, schizophrenia, manic-depressive illness

MINOR SYMPTOMS

1) Neurologic

a) Numbness without objective findings

b) Clumsiness without objective findings

c) Headache

2) Psychiatric

a) Reactive depression

b) Mood swings

c) Severe anxiety and/or other psycho-neuroses

d) Behavioral problems

CSF examination is done to rule out tubercular and fungal infections. CSF pleocytosis, elevated proteins specialy-Gilal fibrillary acidic protein (GFAP) and neurofilament triplet proteins can be found in flare.

Various serologic, electrophysiologic, and neuroimaging findings have been proposed as markers of NPSLE; however, none are diagnostic. Three serologic markers that are regarded as fairly specific for NPSLE include antineuronal, antiribosomal, and antilymphocytotoxic antibodies. Currently, however, only a few centers are capable of performing this test.

However other investigations like low hemoglobin, high ESR, low C3, C4 levels, and rising titers of dsDNA support the diagnosis of flare .

Imaging studies like CT scan to rule out hemorrage, and MRI to rule out any focal or diffuse white matter involvement is also indicated.

The diagnosis of NPSLE, though supported by diagnostic studies, remains a clinical one. Some studies suggest that a post treatment residual cognitive deficit often remains.

Our patient had presented with major psychiatric manifestation i.e. acute onset of psychosis that was precipitated by exaggerated mental stress after her class tenth result, in which she had failed. And the latter could have been the result of her recent cognitive decline. Her dsDNA titers and ESR had increased as compared to the previous reports.

Conclusively, individuals with major manifestations of NP-SLE require aggressive therapy with high dose steroids and perhaps immunosuppressive drugs, plus other appropriate treatment such as anti-seizure medication. Azathioprine is added for maintenance. IVIg is reserved for nonresponders. Anticonvulsants, antipsychotic drugs can be added as per indication.

But what should be done for patients with minor manifestations of neurologic and psychiatric NP-SLE?

 At present, the treatment of symptoms is most judicious, such as the control of headaches and migraines with appropriate medications.

References:

1. Lupus flare ;how to diagnose and treat-Lata Bichile, Vaibhav Chewoolkar,-Medicine update 2011(281- 286)
2. Khoshbin S,Glanz Bl,Schur PH:Neuropsychatric syndromes in SLE, new look,Cli Exp Rheumatol 1999Aug,17(4)-395-8.
3. F.B,Karassa,J.P.A.Ioannidis-Risk factors for CNS involvement in SLE –QJM, vol 93, p-169-174
4. Michelle Petri M D, MPH Mark –Athritis and rhumatism, vol 34 p 937-944
5. Tarakad S Ramachandran, MBBS, FRCP(C), FACP; Chief Editor: B Mark Keegan,MD, FRCPC, Medscape – Neurological Manifestations of SLE updated June 29 2011