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Lupus, Disease of A Hundred Faces

This disease is popularly known as “disease of a hundred faces” due to its manifestations which can mimic around one hundred other diseases and therefore hampers diagnosis. It develops very slowly in the course of many years, with varied signs and symptoms, like pieces of a puzzle which contribute to delay in the diagnosis.

Common symptoms are weakness, daily prolonged tiredness, physical and mental fatigue, low grade fever, anorexia, weight loss, hair loss, sore muscles, joint pain without any arthritis and photo sensitivity which causes facial butterfly rash. Because all these symptoms are similar with other diseases, physicians frequently do not think of lupus the first time.

John Darmawan, MD PhD FACR, a rheumatologist from Semarang who is also WHO Expert on Rheumatic Disease, stated that diagnosis of lupus should fulfilled five out of 11 criteria of American College of Rheumatology. Those five criteria are to be collected base on the disease history.

Some of the criteria are mentioned above, including unspecific symptoms before arthritis signs develop which occur only for a few months, painless mouth ulcer which does not resolve over several weeks, long-lasting facial rash, and photosensitivity to the sun (part exposed to the sun becomes reddish for several hours or longer).

If there is only less than 1-2 criteria, lupus diagnosis could be confirmed with laboratory tests. If one or two laboratory tests are positive, like ANA (antinuclear antibody) test and severe anemia, the lupus diagnosis is confirmed. Lupus could also be indicated by leucocytes less than 4000/cc, thrombocytes less than 100.000/cc etc. Other than blood, kidney and immune function could also be an indicator of lupus.

The low incidence of lupus (40/100,000), make it difficult for physicians to find lupus cases in their practices.

Joint management

According to John Darmawan, usually lupus is treated by a rheumatologist. However, the complexity of lupus disease and its treatment requires cooperative management together with other specialists, depending on which body organ is involved. Kidney lupus is better managed together by nephrologist and rheumatologist; skin lupus together with dermatologist and brain lupus together with neurologist.

In general, there are three kinds of lupus, i.e. SLE (Systemic Lupus Erithematosus), discoid lupus, and drug-induced lupus. Drug induced lupus due to drug adverse reaction will resolve by itself when the drug is stopped. Discoid lupus is a lupus of the skin that manifests as various skin abnormalities.

SLE could cause complications such as brain lupus, lung lupus, digital vascular lupus of the hand and foot, skin lupus, lupus of the kidney, heart lupus, blood lupus, muscle lupus, retinal lupus, joint lupus, etc.

Choice of drug depends on the kind of lupus treated. All drugs, including drug for lupus disease has adverse effects. Since medication is to be taken long-term, adverse effect should be prevented by providing the body with enough calcium and potassium from food, drinks (milk and dairy products), fruits and vitamin D. It is also to prevent bone loss because lupus and lupus drugs could cause bone loss.

Risk factors

Lupus is found in both sexes at all ages. However, risk of lupus in adult woman at reproductive age is eight times higher than adult male.

Sulpha drugs, penicilline, hidralazine, procainamide, and also ultraviolet ray, and infection, can induce lupus in woman with predilection for this disease. Patient in remission with maintenance therapy and in drug-free remission can experience flares if risk factors such as sunlights, physical and mental stress is not avoided.

Year-long sun exposure in tropical countries like Indonesia may become a factor for disease relapse. Patient who is sensitive to the sun can get a facial rash just driving from Magelang to Semarang,” said John Darmawan.

Some things are to be avoided by lupus patients, including direct sunlight, reflection of sunlight from the road into the car or ultraviolet through should be avoided.

Injection with silicone for lips, cheek or breast and buttock enlargement should never be for lupus patients. Hair-dye is prohibited as well,” he said.

Working overtime, laborious physical work, strenuous exercise should be avoided. Lupus patient with diabetes as well as those with history of gastric bleeding, should not take steroids. Joint with acute arthritis condition (swelling, redness, tenderness, pain and stiffness) should not be exercised, except for passive movements that do not trigger pain.

Recommendation

Lupus patients should always be morally supported by his/her closest person because stress could emerge anytime. Regular visits according to the physician’s advice should be done. If there is suspicion of relapse, the attending physician should be contacted immediately. The timeframe between start of relapse and report to the physician should not exceed 7 days.

Lupus patients are also advised to wear clothes covering arms and legs, wear a hat or umbrella impenetrable by ultraviolet rays when going out.

Daily menu recommended by John Darmawan for lupus patients is adequate nutrition rich in calcium, potassium, zinc, vitamin B6, C and D. Patients are also advised to eat a high protein, low carbohydrate diet.

All fruits and vegetable are recommended. For example, bananas, melon, dried fruit, *pisang sale* (caramelized banana), jackfruit, durian, asparagus, broccoli, edible roots, spinach, waterspinach, etc. Milk, yoghurt and cheese are also recommended. (SN Wargatjie).

Striving for remission without therapy

Five-year survival rate for lupus (SLE) patients in Indonesia at this moment is 100% and 85.5% of that can attained drug-free remission for more than 10 years.

According to John Darmawan, MD, PhD FACR, WHO Expert on Rheumatic Disease, more than half a century ago, lupus is still a fatal disease. In the 1950's five-year survival rate of lupus patients was only 50% in western countries and 13% in developing countries.

In a report by Prof Handono Kalim and Kusworini Kalim from Malang, five-year survival rate reached 68% in 1996. While Drenkard et al. (Baltimore, USA) in 1996 reported that 23.4% out of 667 lupus patients, including patients with severe complications, attained drug-free remission.

John Darmawan in his report (Indonesian Medical Journal volume 49, no. 5, May 1999) stated that out of 62 patients, 85.5% attained drug-free remission and five-years survival rate was 100%.

This report was based on the observation of 62 lupus patients under his care. They came from different cities in Indonesia and 21 patients were referred from abroad with severe complications and illness duration of two to five years.

Improvement of five-years survival rate and drug-free remission within 10 years clearly brings a new hope for patients and their family. This success was also reported by John Darmawan in international scientific journals, i.e. APLAR Journal of Rheumatology (vol.3 no. 2, September 1999) and JAMA SEA (March/April 2000 edition) as well as in Indonesian Medical Journal (vol. 49, no. 5, May 1999).

John Darmawan stated that improved survival was achieved with the right medication selection, the right dose and the right methods of administration. This will help to speed up attaining remission with maintenance therapy and then to drug-free remission. "The availability of antibiotics and early eradication of infection lately is very crucial to prevent death," he said.

Millenium therapy of lupus

John Darmawan explained that millenium therapy of lupus consisted of low-dose immunosuppressants combination given intravenously, a mixture of methylprednisolone, cyclophosphamide, methotrexate and oral cyclosporine with mycophenolate mofetil.

After remission in maintenance therapy is attained, intravenous infusion is gradually stopped and replaced by oral methotrexate with cyclosporine and mycophenolate mofetil until drug-free remission is achieved.

"There are many variation of immunosuppressants combination in term of dosing, methods of administration and frequency to achieve the maximum result," said John Darmawan.

Combination of hydroxychloroquine and prednisone is first drug of choice in SLE without complications. This combination is cheap, can be administered by general practitioner and is safe to be used during pregnancy.

SLE with complication is an emergency and need immediate treatment with immunosuppressant combination therapy. Therapy with one immunosuppressant in high dose is not adequate and is prone to adverse effect like facial edema, growth cessation (in child patients) etc.

Generally, SLE without complications can attained drug-free remission after two to four years of treatment, depending on how early treatment was started. Earlier treatment of SLE will achieve remission with maintenance therapy faster and then moving to drug-free remission.

End result of therapy depends very much on the disease activities, which can be monitored by erythrocyte sedimentation rate (ESR) in 1 hour and dilution of C-Reactive Protein (CRP). By maintaining both laboratory factors within normal limit as long as possible, drug-free remission and remission with maintenance therapy could prevent fatal complication and early death.

Smooth and fast communication by e-mail, telephone, and facsimile whenever early signs and symptoms, drug adverse events and infection appears is crucial to the duration of remission with maintenance therapy and drug-free remission.

In developing countries, socioeconomic factors also determine the survival of lupus patients. Underdeveloped health insurance and social security system make it difficult for patients from middle-low economic status to pay for optimal treatment.

Unavailability of continuous payment for treatment causes inadequate therapy, which could trigger complication that generally lead to death.

Lupus patients at early stage need Rp. 5 million to achieve remission with maintenance therapy and around Rp. 10 million to attain drug-free remission. For lupus patients with complications, the cost would be higher.

Lupus in pregnancy

John Darmawan stated that women with lupus in remission using maintenance therapy and in drug-free remission may get pregnant and deliver healthy babies. However, oral methotrexate and oral cyclophosphamide should be replaced with methylprednisolone, or cyclosporine, or mycophenolate mofetil.

During pregnancy and post-delivery, the patient should be monitored closely by her physician and obstetrician to prevent a flare during pregnancy and after delivery. There is a tendency for lupus to relapse in the second and third semester after giving birth. "There is no problem if a mother in remission using maintenance therapy is going to nurse her baby," he said.

(SN Wargatjie).

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