Avascular necrosis of the right femoral head in a systemic lupus erythematosus patient

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According to the 1993 Association Research Circulation Osseous, idiopathic avascular necrosis of the femoral head is defined as the presence of disease or other causes that result in ischemic osteonecrosis of the femoral head without the presence of trauma or sepsis. Based on the above definition, idiopathic avascular necrosis (AVN) includes those that are the result of steroid administration, systemic lupus erythematosus, alcoholic consumption, etc. The pathogenesis of AVN is still obscure; however, it is basically caused by vascular circulation disorder, cell death and decreased capability of bone repair.\(^{1,2}\)

Systemic lupus erythematosus (SLE) is characterized by the presence of systemic immune dysregulation, autoantibody formation, immune complex in the circulation, and activation of the systemic complement. The pathology during recurrence of SLE, among others, is the presence of vascular lesion in the form of inflammation, thrombosis, endothelial injury in which the three of them are predispositions for atherosclerosis. The vascular lesion will cause microcirculation damage which is a risk factor for the occurrence of AVN in activation of SLE. Besides being caused by vascular lesion during activation of SLE, AVN is also triggered by fat deposition in SLE patients as a result of long term steroid therapy that causes abnormal blood fat level.\(^{1,2}\)

Patients with SLE who have undergone pharmacologic treatment with systemic steroid either in oral or injection form will have 10 to 40 times the risk of having idiopathic AVN. High dose of systemic steroid treatment of more than 4000 mg of prednisone administered for more than three months or low dose of oral steroid administered for seven days can become a risk factor for AVN. The mechanism of AVN caused by steroid treatment is associated with hypercoagulation, fibrinolysis disorder, and thrombosis of the bone vein.\(^{1,2,3}\) We report a case of AVN of the right femoral head in an SLE patient.

CASE REPORT

A 29 year-old male has been diagnosed with SLE since 2006. During the last five months of the follow up, the patient complained of pain of the right hip which worsened when walking and therefore causing a slight limp. The patient also complained of reddening of the face when exposed to sunlight and red spots appearing on the skin of the palms and the abdominal area. The results of physical examination were as follows: mental state was compos mentis, blood pressure was 120/70 mm Hg, pulse rate was 84 times/minute, respiratory rate was 16 times/minute, and body temperature was 36.5°C. The conjunctiva was not pale. There was no abnormality detected in ears, nose, and throat. On lung examination, the main respiratory sound was vesicular breathing in both lungs and there was no additional sound. On cardiologic examination, the heart border was normal, S1 and S2 were regular, and no murmur was detected. On abdominal examination, the abdomen was supple on palpation, the liver and spleen were not palpable, intestinal sound was normal on auscultation. Malar rash was seen on the facial skin while vasculitis was seen on the skin of the palmar area, thoracic area, upper abdominal area, volar upper arms, and lower extremities.

Figure 1 Malar rash of facial skin

Figure 2 Vasculitis of the thoracic and abdominal skin
The results of the blood test were as follows: Hb level was 16.7 g/dl, leukocyte count was 9,200/ul, platelet count was 232,000/ul, SGOT level was 33 U/L, SGPT level was 32 U/L, gamma GT was 26 U/L, serum alkaline phosphatase level was 183 U/L, BUN level was 20 mg/dl, serum creatinine level was 1.0 mg/dl, BSN/2JPP: 72/85 mg/dl, total cholesterol level was 152 mg/dl, triglyceride level was 120 mg/dl, HDL cholesterol level was 30 mg/dl, LDL cholesterol level was 97 mg/dl, Na level was 137 mEq/L, K level was 3.46 mEq/L.

The diagnoses established were SLE with involvement of the skin and vasculitis, and AVN of the right femoral head. The patient was then treated with 50 mg once per day of azathioprine, 4 mg twice per day of methyl prednisolone, 500mg three times per day of CaCO₃, 35 mg once per day of risedronate, and 80 mg once per day of acetylsalicylic acid. A total hip arthroplasty was later performed and resulted well.

**DISCUSSION**

Avascular necrosis is a disease that can occur in SLE. The incidence is between 5 to 80 percent. The risk factors of AVN, among others, are: steroid consumption either in high dose or in low dose administered for more than seven days, age, vasculitis, Raynaud’s phenomenon, thrombophlebitis, the presence of cardiopoline antibody, and increased activity of SLE. Glucocorticoid therapy is associated with the occurrence of AVN through the following mechanism: blood stasis, trabecular bone ischemia, and increased osteocyte apoptosis. It is still obscure whether the mechanism of AVN resulting from glucocorticoid therapy is associated with the formation of thrombosis and fat emboli. Oinuma et al reported that the onset of AVN in SLE patients undergoing a high dose of glucocorticoid therapy occurs usually during the first month of therapy. The patient consumed a low dose of steroid routinely in order to control the SLE; however, the disease had become active since five months prior to the detection of AVN with signs of recurrence of malar rash and vasculitis of the skin. In addition, there was another risk factor, that is, the patient experienced dyslipidemia prior to treatment using the statin group. The main pathogenesis of AVN is vascular injury and fat deposition. Vascular injury is manifested by vasculitis and probably fat deposition because the patient had experienced dyslipidemia.

In order to establish the diagnosis of AVN, besides using clinical signs such as tenderness of the avascular necrotic bone area, radiographic or MRI examination should be performed. The laboratory examination result can not be used as a basis for the diagnosis. Based on the modified Steinberg classification, AVN of the femoral head is classified into the following stages/grades:

1. Radiographic result is normal but MRI or bone scan shows mild to severe abnormality of the femoral head,
2. Radiographic result shows mild to severe changes of the femoral head which became radiolucent and sclerotic,
3. Radiographic result shows subchondral fracture-Crescent sign–and mild to severe abnormality of the femoral head,
4. Radiographic result shows subchondral fracture and levelling of the femoral head with mild to severe involvement of the femoral head,
5. Radiographic result shows narrowing of the joint and/or changes of the acetabulum,
6. Radiographic result shows severe degeneration

Involvement of the femoral head is mild if the percentage of involvement is 1 to 25%, moderate if it is 26 to 50%, or severe if it is more than 50%. Based on the above classification, we can conclude that the patient had stage/grade 4 AVN of the femoral head.
The management of AVN depends on age, AVN stage/grade, occupation, and prior treatment. Basically, the management of AVN is to avoid the triggering factor by improving fat metabolism and blood circulation, to give treatment to prevent deterioration of the SLE, and to undergo surgery. The surgery performed - core decompression, valgus osteotomy, bone grafting - is for the mild or early stage/grade while a total hip arthroplasty is performed for the severe and late stage/grade.\textsuperscript{1,2,6-10} The patient is still given a low dose of steroid but in a much lower dose and then combined with azathioprine, statin therapy for managing dyslipidemia, acetylsalicylic acid, biphosphonate, and total hip arthroplasty.

**SUMMARY**

We report a 29 year-old SLE male patient who had complained of tenderness of the hip and right foot which caused a limp when walking, reddening of the facial skin, and appearance of red spots on the chest and abdominal skin since five months ago. The patient was finally diagnosed with activated SLE with involvement of the skin and vasculitis, and AVN of the right femoral head. In addition to continuing the therapy to manage the SLE, a total hip arthroplasty was performed to treat AVN of the femoral head.

**REFERENCES**