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THE BONE THIEF: MULTIFOCAL OSTEONECROSIS IN JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS

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INTRODUCTION

Osteonecrosis (ON) is a common form of skeletal damage seen in Systemic Lupus Erythematosus (SLE) together with avascular necrosis (AVN) and fragility fractures . ON is often attributed to long term use and high cumulative corticosteroid (CS) doses which promotes conversion of red marrow in bone to fat, resulting in elevated marrow pressure, impaired perfusion and ultimately bone cell death. Here we highlight two teenage girls with juvenile SLE and extensive multifocal atraumatic ON of the lower limbs due to multiple contributing factors.

CASE 1

This 16-year old Chinese girl first presented at age 13 years with severe SLE (SLEDAI score 24) & Lupus Nephritis Class IV (A) with renal impairment. She required intermittent haemodialysis, CS and IV Cyclophosphamide (CYC) with good initial response but was complicated with an episode of microangiopathic hemolytic anemia 6 months later, requiring Rituximab. She achieved complete renal remission with 12 monthly doses of IV CYC and her Prednisolone weaned. She unfortunately developed right hip AVN at 14.5 years whilst on low dose Prednisolone 5 mg OD and Mycophenolate mofetil total 1.5 gram OD. Unsatisfied with the conservative approach adopted for AVN, she sought oral traditional treatment and reappeared two months later looking severely Cushingnoid with florid right knee synovitis and pain on weight bearing. She responded to empirical treatment for septic arthritis (5 weeks of IV Imipenem) with an arthroscopic washout which yielded sterile synovial fluid (both bacteria and tuberculosis). Three months later, she developed recurrent left knee synovitis which transiently responded to intra-articular CS injections but subsequently remained refractory to oral CS and subcutaneous Methotrexate. MRI left lower limb showed extensive osteonecrosis of the femur and tibia extending to the talar dome with osteochondral defects and fragments within the knee joint (images A & B). Arthroscopic debridement and synovectomy of the left knee was performed and the arthritis resolved thereafter.

CASE 2

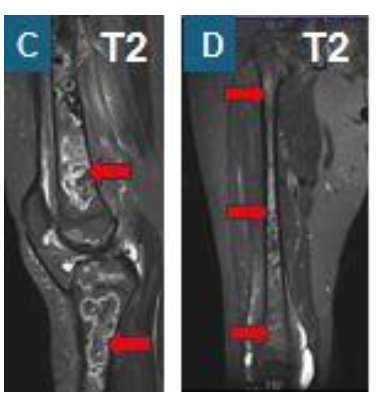
This 18-year old Malay girl first presented at age 11 years with severe multiorgan SLE (SLEDAI score 26). She responded initially to CS, Hydroxychloroquine, and Azathioprine. However, due to challenging social circumstances, her compliance to treatment and follow-up was poor. Her first renal flare was at age 13 years when she had Lupus Nephritis Class III and was treated with 6 months of IV CYC. Concurrently, she complained of back pain and sustained an L1 osteoporotic vertebral fracture treated with IV Pamidronate 3 monthly for 2 years. Her second renal flare was at age 17 years whilst on Azathioprine maintenance whereby repeat renal biopsy showed Lupus Nephritis Class IV (A/C) + V. During the first cycle of CYC, she developed right knee *Streptococcal agalactiae* septic arthritis and completed 6 weeks Cefuroxime. She responded well and continued with monthly IV CYC and Rituximab, and achieved renal remission by the 8th month. Unfortunately 3 weeks after her 11th CYC cycle, she had recurrence of right knee arthritis which was treated as septic origin (elevated CRP 8.79mg/dL (normal <0.5) and procalcitonin 11.2µg/L (normal <0.05) with 3 weeks of IV Ceftriaxone and 3 weeks of PO Clindamycin. Her synovial fluid culture was sterile and she improved clinically with normalization of CRP after 3 weeks of antibiotics. However, her right knee arthritis recurred at 5th week of antibiotics and a joint aspirate was similarly sterile prompting an MRI which showed multifocal osteonecrosis affecting the entire femur, tibia, and around the knee (images C & D). The knee swelling subsequently resolved spontaneously.

A:
Osteochondral defect (⇨) at knee joint

B:
Osteonecrosis (⇦) at distal tibia extending to talar dome (⇧).

C:
Osteonecrosis (⇦) around knee joint

D:
Osteonecrosis (⇨) of entire femur



DISCUSSION & CONCLUSION

SLE patients have a higher risk of developing ON compared to other CS-treated rheumatic diseases. Apart from the contribution of CS, additional risk factors in our patients included the presence of high disease activity, vasculopathy, adolescent age at diagnosis and treatment escalation during flare-ups. Other risk factors for ON are antiphospholipid syndrome, Raynaud's phenomenon, and genetic susceptibility. One must also not forget the contribution of non-prescription CS (traditional medications) as in Case 1 and infection in Case 2. We highlight the severe extent of ON that can occur in SLE and how reactive synovitis from bony or cartilage fragments could incite an intense inflammatory response and mimic a SLE arthritis or septic arthritis. Therefore, clinicians should consider ON as a cause of recalcitrant synovitis in any SLE patient, especially those with multiple risk factors.

References
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