(*Refer to page 172*)

Answer: SLE (Systemic lupus erythematous)

From epidemiological studies, SLE is rare in males (M: F ratio of 4.7:44) but is more common in Asian and African population compared to Caucasians.[1] There is no reported prevalence of SLE amongst Bruneians. However, from an Asian study, the prevalence of SLE in Asian countries has been reported to be variable, from 3.2-19.3/100,000 population in India, Japan and Saudi Arabia, and up to 70/100,000 recorded in Shanghai.[2]

Systemic lupus erythematous (SLE) is the archetypical autoimmune disease of an unknown aetiology and present with a diverse spectrum of phenotypical features involving almost any organ system. There are several predisposing factors including heredity, complex genetics, complement deficiency and sex hormone status. [3] Azizah and colleagues found out that HLA- DR2, DQB1*0501 and DQB1*0601 are the most common HLA antigens present in Malay patients which are associated with a higher incidence of malar rash, mouth ulcers and lupus nephritis but not a younger age of disease onset. [4]

The reasons supporting early onset of diagnosis as is with our case are still relatively unknown and are most likely due to genetic and environmental factors supported by a strong and complex genetic predisposition with a high concordance rate among monozygotic twins and familial aggregation amongst relatives of people with SLE.[5] A review of the worldwide literature found that the incidence of pediatric-onset SLE ranged from 0.36 to 2.5 per 100,000 per year and the prevalence ranged from 1.89 to 25.7 per 100,000.[6]

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Overall, having an affected first-degree relative with SLE was associated with an adjusted RR (95% CI) of 16.92 (15.23-18.80) sibling risk ratio 8-fold to 29-fold higher than that in the general population and a 10-fold increase in disease concordance in identical twins. In addition, there is a 24-56% concordance rate in monozygotic twins, compared with a 2-5% risk in dizygotic twins.[7]

Treatment is based on the severity of end-organ involvement and induction therapy is usually commenced with corticosteroids (or IV cyclosporine or cyclophosphamide for life threatening disease or lupus nephritis) followed by maintenance treatment with a steroid sparing agent (such as azathioprine, methotrexate and mycophenolate) aiming to reduce organ failure and mortality. [1] Additionally, NSAIDS and/or hydroxychloroquine (HCQ) is routinely used in most SLE patients for the reduction of arthralgia and constitutional symptoms.

The prognosis of SLE has improved significantly with reported average 10-yr survival rate exceeding 90%, with good medical management. Mortality was attributed to the disease itself in contrast to the present data that mortality is often a result of medication side effects or cardiovascular events. Hence, these patients should be managed closely with proper titration of their medical therapy.

REFERENCES

1: Professor C. A. Inderjeeth, Habib P., Chanakya S. et. al. SLE- Reducing life threatening progression MedicineToday 2018: 19(9): 31-41.
2: E Osio- Salido and H Manapat- Reyes Epidemiology of SLE in Asia Lupus (2010) 19, 1365–1373.
3: Rhodes B, Vyse TJ. The genetics of SLE: an update in the light of genome-wide association studies. Rheumatology (Oxford) 2008: 47: 1603- 1611..
4: MR Azizah, SS Ainol, NCT Kong et. al. HLA Antigens in Malay Patients with SLE: Association with Clinical and Autoantibody Expression. The Korean Journal of Internal Medicine Vol. 16, No. 2, June 2001, 123-131 .
5: Kuo C- F., Grainge M., Valdes Ana. et. al Familial Aggregation of Systemic Lupus Erythematosis and Coaggregation of Autoimmune Disease in Affected Families JAMA Intern Med. 2015; 175(9):1518-1526 .
6: Pineles D; Valente A; Warren B; Peterson MG; Lehman TJ; Moorthy LN. Worldwide incidence and prevalence of pediatric onset systemic lupus erythematosus. Lupus. 2011; 20(11):1187-92 (ISSN: 1477-0962) .
7: Yun Deng and Betty P. Tsao. Genetic susceptibility to systemic lupus erythematosus in the genomic era Nat Rev Rheumatol. 2010 Dec; 6(12): 683–692.