E-Poster Presentation

1. Antiphospholipid syndrome

LP-001 ANTIPHOSPHOLIPID SYNDROME IN JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS: THE MALAYSIAN EXPERIENCE

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Background Juvenile onset Systemic Lupus Erythematosus (jSLE) is a rare multisystem autoimmune disease with variable clinical features and severity. jSLE is a common cause of secondary antiphospholipid syndrome (APS). Our aim is to describe the clinical characteristics, treatment, and outcomes of jSLE with secondary APS in our cohort.

Methods A retrospective descriptive study of all jSLE patients with APS from January 2013- December 2022 (10 years) seen at the Paediatric Rheumatology Unit, Selayang Hospital, Malaysia. Clinical, laboratory, treatment and outcome data were collated from hospital electronic medical records.

Results We identified 11 patients with total of 14 thrombotic events. The mean age of SLE diagnosis: 11.0 years (8.9–14.9) and mean age of first APS diagnosis: 13.9 years (10.2–21.3). The majority were Malays (36%) and Indians (36%) and the female-to-male ratio was 10:1. Most were venous thrombosis (79%) predominantly lower limb deep vein thrombosis (73%). Arterial thrombosis occurred in 14%, affecting the internal carotid and central retinal artery; while one (7%) had a mixed arterial (splenic infarct) and venous thrombosis (bilateral pulmonary artery). Of those tested, positive antiphospholipid antibodies include lupus anticoagulant (80%), beta 2-glycoprotein (56%) and anticardiolipin antibody (22%). Risk factors included obesity (4/11), overweight (4/11), hypertension (3/11) and high cholesterol (3/11). The mean SLEDAI score was 7 at time of thrombosis (range 0–22). Eight patients had a prior diagnosis of jSLE and were receiving Prednisolone (100%), Hydroxychloroquine (75%), Mycophenolate mofetil (38%), Azathioprine (25%) or Cyclophosphamide (13%). All received heparin during the acute phase (10/11 sc Enoxaparin) followed by warfarin. One required surgical intervention (transcatheter embolectomy and inferior vena cava filter insertion). All patients survived with complete clinical resolution of thrombotic episodes.

Conclusions Antiphospholipid-associated thrombosis in juvenile SLE is predominantly venous, sometimes recurrent and occurs during active SLE disease. Unlike adults, traditional risk factors are uncommon in jSLE and the overall prognosis is good.

LP-003 ANTIPHOSPHOLIPID ANTIBODIES AND RECURRENT THROMBOSIS IN PRIMARY ANTIPHOSPHOLIPID SYNDROME

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Background Antiphospholipid syndrome (APS) is acquired thrombophilia characterized by the presence of antiphospholipid antibodies (aPL). Its main manifestations are vascular thromboses and pregnancy complications. Despite apparently adequate anticoagulation, the risk of recurrent thrombosis remains high.

The aim. To evaluate the relationship between aPL and recurrent thrombosis in patients with primary APS (PAPS).

Methods The study included 52 patients with PAPS: 30 (58%) – women and 22 (42%) – men. The mean age of the patients was 38.5 [31.5–43.5] years, duration of the disease – 9.0 [3.1–13.0] years. Recurrent thromboses had 34 (65%)/52 patients and no recurrent thromboses had 18 (35%)/52. The study of aPL involved the determination of IgG/IgM antibodies to cardiolipin (aCL), IgG/IgM antibodies to β2 glycoprotein 1 (anti-β2GP1) and IgG/IgM antibodies to phosphatidylserine-prothrombin complex (aPs/Pt) by enzyme immunoassay (ELISA), IgG/IgM/IgA aCL, IgG/IgM/IgA anti-β 2GP1, IgG antibodies to domain I β2 glycoprotein 1 (IgG anti-β2GP1DI) by chemiluminescence assay (CLA).

Results IgM aCL and IgM anti-β2GP1 were not associated with recurrent thrombosis, in contrast to IgG aPL (ELISA/CLA); figure 1. There was an association between recurrent thrombosis and «extra»-criteria aPLs: IgG aPs/Pt, IgA aCL, IgG anti-β2GP1DI.

Combination of IgG aCL+IgG anti-β2GP1+IgG aPs/Pt (ELISA) was 34/52 (65%) patients and thrombosis recurred in 27 (79%)/34; p=0.008. Thirty-six (69%)/48 patients had a combination of IgG aCL+IgG anti-β2GP1+IgG anti-β2GP1DI (CLA). Recurrent thromboses had 29 (81%)/36; p=0.0003. . Relationship between antiphospholipid antibodies and recurrent thrombosis in patients with primary antiphospholipid syndrome is shown in table 1.

Conclusions Recurrent thrombosis was associated with triple IgG aPL positivity in any combination Patients with positive IgG aPL (ELISA/CLA) and IgA aCL were significantly more likely to have recurrent thrombosis. The highest relationship was observed with IgG anti-β2GP1DI (CLA) and IgG anti-β 2GP1.