decided to study the incidence and clinical spectrum of APS in a general hospital.

Objectives To examine the spectrum of clinical manifestations of antiphospholipid antibody syndrome (APS) in the acute general hospital of the southern area of Israel.

Methods Clinical records of patients who were discharged with diagnosis of APS during the years 1990–2000 were reviewed.

Results A total of 22 patients fulfilled criteria of APS (accordingly to Asherson's criteria, 1988) with 38 clinical events. Primary APS was found in 15 patients and secondary in 7 patients. There were 19 female and 3 male, mean age was 42.1 years (from 29 to 56). Most of the patients (20 out of 22) were diagnosed after 1995, showing increased awareness of this syndrome among the staff.

16 thrombotic events occurred: 6 stroke (27%), 4 acute myocardial infarction (18%), 3 venous thromboembolic event (13.6%), 3 recurrent abortion (13.6%). 32 nonthrombotic events were found: 8 headache (36.3%), 6 thrombocytopenia (27%), 3 skin rashes (13.6%), 1 amaurosis fugax (4.5%), 1 catastrophic APS (4.5%) 1 progressive supranuclear palsy (PSP) (4.5%).

All patients suffering from acute myocardial infarction were patients with SLE, thus showing that APS is an additional risk factor for ischaemic heart disease in such patients.

Headache was an additional symptom in the patients, diagnosed as having APS – in all 6 patients with stroke, one with amaurosis fugax and one with venous thromboembolic event.

Patient with PSP features had magnetic resonance imaging of brain and blood test results characteristic to APS (our unpublished data).

Conclusion The study shows that APS is not an uncommon disease. It is important for the internist and the specialist to be aware of the clinical manifestations of APS. Its inclusion in the differential diagnosis of arterial or venous thrombosis with or without thrombocytopenia in patients is obligatory at the present time.

FR10178 FOLLOWUP OF PLATELET ACTIVATION IN A PATIENT WITH ANTIPHOSPHOLIPID SYNDROME UPON TIROFIBAN TREATMENT

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Background Patients with primary or secondary antiphospholipid syndrome (APS) often suffer from stroke leading to high degree of disability. Though it is well known that phospholipid antibodies induce a procoagulatory scenario, antibody status does not well characterise patients at risk. Using transcranial Doppler ultrasound of intracerebral arteries we could detect microembolic signals correlating well with phospholipid antibody titer as well as with history of stroke. In these patients we could also demonstrate elevated levels of platelet activation markers (p-selectin) as well as increased platelet leukocyte aggregation. Tirofiban is a potent, highly selective and reversible inhibitor of platelet gpIIb/IIIa receptor which binds fibrinogen on platelet surface.

Objectives We treated a patient with primary APS and refractory transitory ischaemic attacks with tirofiban and detected microembolic signals as well as platelet activation markers and platelet-leukocyte aggregations (PLA).

Methods In fresh venous citrate-anticoagulated whole blood expression of gp Ib (CD41), gp IIb/IIIa (CD42b), fibrinogen binding and p-selectin (CD62p) was analysed using a flowcytometric double staining method. In parallel we detected PLA by determining percentage of platelet marker/leukocyte marker (CD42b/CD45) double positive leukocytes.

Results Upon Tirofiban treatment we found a reduction of platelet activation markers (p-selectin, fibrinogen binding, CD63). Under steady state conditions with tirofiban expression of platelet activation markers and platelet leukocyte interaction normalised completely compared to normal healthy donors. The morning after each infusion there was a rebound phenomenon, which could also be observed in platelet activation markers and PLA. This rebound phenomenon could be reproduced upon sequential administration of tirofiban in parallel with rate of microembolic events.

Conclusion In a patient with primary APS therapeutic intervention with tirofiban, a reversible inhibitor of platelet gpIIb/IIIa receptor, induced a clinical improvement of transitory ischaemic attacks. In parallel with clinical improvement there was a modulation of microembolic signals as well as platelet activation markers and PLA. Flowcytometric measurement of platelet activation can be a useful tool in monitoring platelet hyperreactivity in APS upon therapeutic intervention.

FR10179 NAILFOLD CAPILLAROSCOPY IN SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS WITH AND WITHOUT ANTIPHOSPHOLIPID SYNDROME

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Background Nailfold capillaroscopy is able to evaluate microcirculatory damage in many different diseases and a high prevalence of capillary abnormalities has been reported in systemic lupus erythematosus (SLE).

Objectives Considering that the vascular involvement is of primary importance in antiphospholipid syndrome (APS), it can be of interest to examine the capillaroscopic features of those SLE patients having secondary APS.

Methods 29 SLE patients entered the study, 13 had secondary APS (M/F = 1/12; mean age = 45.5 yrs; mean disease duration = 5.85 yrs) while the other 16 patients (M/F = 1/15; mean age = 36.6 yrs; mean disease duration = 7.2 yrs) had no clinical or laboratory signs of APS. There were no significant differences regarding the number of ARA criteria or ECLAM and SLEDAI scores between the two groups.

Each patient underwent nailfold capillaroscopy to detect the following abnormalities: morphological alterations of capillaries, variability of capillary loop length, presence of microhaemorrhages, irregular distribution of capillaries and sludging of blood. Moreover to score these changes a semiquantitative rating scale was adopted, according to previous studies.

Results Although not statistically significant, more frequent variability of capillary loop length (77% vs 44%), presence of microhaemorrhages (23% vs 12.5%), sludging of blood (23% vs 12.5%) as well as capillaroscopic score >1 (77% vs 44%) were found in patients with SLE/APS respect to those with SLE alone.

However in our SLE/APS patients the presence of capillaroscopic microhaemorrhages was significantly associated to thrombocytopenia ($p < 0.034$) and the finding of shorter capillary

loop length was related to the occurrence of livedo reticularis ($p < 0.05$).

Conclusion Thus we suggest that nailfold capillaroscopy can be useful to better define SLE patients with APS, helping to identify some microvascular changes that seem related to clinical and laboratory features of APS.

FRI0180 PARANEOPLASIC ANTIPHOSPHOLIPID SYNDROME- A POSSIBLE MAIN CAUSE OF THROMBOSIS IN CANCER PATIENTS

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Background The antiphospholipid (APS) is a hypercoagulable state marked by the antibodies that recognise anionic phospholipid-protein cofactor complexes.

Objectives This study was undertaken to investigate a possible association of antiphospholipid antibodies (APLA) in cancer patients with thromboembolic events.

Methods The study group included 46 patients with different malignancies and associated thromboses. Tests were performed for detecting APLA: anticardiolipin enzyme linked immunoassay (ELISA) for both IgG and IgM isotypes, anti-β₂-GPI IgG antibody ELISA, and coagulation test for lupus anticoagulant (LAC). We used ACL method with IL Test LAC Screen and IL Test LAC Confirm.

All patients underwent complete clinical and radiological examination, routine laboratory tests were done.

Results 18 patients had APS associated with evolutive cancers. LAC was present in 14 patients (B-cell lymphoma (2 cases), ovarian-(3), breast-(3), lung-(4), pancreatic-(1), bladder (1) cancer, and was related mainly with deep vein thromboses of the legs, pulmonary embolism and catastrophic antiphospholipid syndrome. Anticardiolipin antibodies (ACLA) were detected in 3 patients with bladder -, pancreatic - and breast cancer, and were associated with aortic nonocclusive thrombosis, stroke, myocardial infarction and multiinfarct dementia. In one patient (prostatic cancer) were present both ACLA and LAC, associated with stroke, myocardial infarction, multiinfarct dementia. The presence of LAC seems to be related more to venous thromboses and metastatic disease, while ACLA seems to be related to arterial thromboses.

Conclusion The presence of APS in cancer patients is associated with vascular thromboses, leading to a poor quality of life or to fatal events (including metastatic spread).

The presence of APLA in cancer patients may be a warning sign of a thrombotic event and might lead to a better selection of patients for prophylactic anticoagulant therapy.

FRI0181 SKIN MANIFESTATIONS IN SECONDARY ANTIPHOSPHOLIPID SYNDROME – ANALYSIS OF FORTY PATIENTS

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Background Skin lesions are common kind of connective tissue disease manifestations.

Objectives The aim of study was evaluation of skin involvement in clinical presentation of 40 patients with systemic lupus erythematosus (SLE) and secondary antiphospholipid syndrome (APS).

Methods Examined group (34 women and 6 men) was represented by patients who fulfilled diagnostic ARA criteria for both, SLE and APS. Average duration of disease was 7.54 ± 7.53 (1–33) years and average age of patients was 31.45 ± 13.95 (8–65) years. Patients with SLE without APS represented control group.

Results We noticed cutaneous ulcerations in 15/40 (37.5%) patients from examined group. They were predominantly located at lower legs and fingers of palms. Cutaneous ulcerations were significantly more frequently ($p = 0.025$) appeared in patients with both, SLE and APS than in control group. Livedo reticularis was observed in 9/40 (22.5%) patients from examined group, the most frequently at lower legs but one patient expressed livedo reticularis at skin of the torso. There was no significant difference in comparison to control group where prevalence of livedo reticularis was 10% (2/20). We observed palmar-plantar erythema in 12.5% (5/40) patients with APS as a part of SLE. In control group, palmar-plantar erythema was noticed in 10% patients (2/20).

Conclusion Our study demonstrated that cutaneous ulcerations occurs more frequently in patients with both, SLE and APS than in patients with SLE alone, probably because of two present pathogenic mechanisms: vasculitis and thrombophilia as a part of APS. On the other hand, livedo reticularis and palmar-plantar erythema were presented with the same frequency in both groups of patients.

FRI0182 CATASTROPHIC ANTIPHOSPHOLIPID ANTIBODY SYNDROME – REPORT OF FIVE CASES

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Background The catastrophic antiphospholipid antibody syndrome (CAPS) is an uncommon disorder, characterised by widespread micro and macrovascular changes due to intravascular thrombosis. We report five cases of CAPS occurred at our department over the past 7 years.

Objectives

Methods All patients were females between 13 and 49 years, all having SLE; two of them had no identifiable precipitant factor, two underwent major surgery, and the last one had a respiratory tract infection and a SLE flare as probable precipitant events; four women had deep vein thrombosis and the other one a peripheral arterial thrombosis; stroke occurred in 4 patients, one of them concomitantly in the right and left middle cerebral arteries territories; intraalveolar haemorrhage occurred in two cases and one patient developed ARDS.

Treatment options were different for each patient and all had steroids and anticoagulation. Two had cyclophosphamide synchronised with plasmapheresis.

Results Unfortunately 4 of the patients died, the only survivor was treated with steroids and anticoagulation therapy.

Conclusion CAPS is a rare event in our cohort of patients with antiphospholipid antibody syndrome (6.9%), having a high mortality rate. Most of the times it was possible to identify a