Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2019-eular.6610

AB0571 **BENEFICIAL EFFECTS OF VACCINATION ON REDUCING RISKS OF INFLUENZA INFECTION IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS**

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Background: In patients with systemic lupus erythematosus (SLE), infection is a major cause of morbidity and mortality. Influenza is one of the most common infectious diseases which can be prevented in a certain degree by vaccination, while in fact, influenza vaccination rates of SLE are really low in China.
Objectives: To investigate the influenza vaccination rate of SLE patients and the reasons for nonadherence to vaccination. To evaluate beneficial effects and safety of influenza vaccination.
Methods: A cross section study was performed among patients with SLE regular follow-up in Distinct HealthCare between June 1 and November 31. Vaccination status and influenza infection condition were surveyed. Demographic information, clinical features and laboratory characters were collected and systemic lupus erythematosus disease activity index (SLEDAI) was documented.
Results: 109 SLE patients were recruited, including 42 immunized with trivalent or quadrivalent split virion influenza vaccine and 67 non-vaccinated. There were no significant differences in demographic and clinical characteristics (p>0.05). The influenza vaccination rate was 38.5%. Influenza infection rates in the vaccinated and non-vaccinated were 7.1% (3/42) and 23.9% (16/67), respectively, with statistic difference (p<0.05). Reasons that non-vaccinated patients reported for nonadherence included refused by the community health workers due to basic disease of SLE (32.3%), vaccine shortages (27.7%), concerns for potential side effects (23.1%) and insufficient patient education (16.9%). No systemic adverse reactions were observed and no significant increase of disease activity was found in vaccinated patients.
Conclusion:
- 1. Split virion influenza vaccine is effective in reducing risks of influenza infection in patients with SLE, and also safe.
- 2. Influenza vaccination is insufficient in SLE patients.
- 3. Education of the community health workers and patients is important to increase influenza vaccination rate.

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Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2019-eular.2756

AB0572 **INVASIVE MYCOSES IN PATIENTS WITH CONNECTIVE TISSUE DISEASE FROM SOUTHERN CHINA: CLINICAL FEATURES AND ASSOCIATED FACTORS**

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Background: Invasive fungal disease (IFD) was well studied in patients with AIDS and organ transplant recipients. A few researches illustrated that patients with connective tissue disease (CTD) were also predisposed to IFD. However, few researches were designed to focus on invasive mycosis (IM) in patients with CTD.
Objectives: To investigate the clinical features and associated factors of IM in patients with CTD from Southern China.
Methods: A retrospective study CTD was performed. Demographic and clinical data were recorded. Associated factors were analyzed by logistic regression analysis.
Results: A total of 32 patients with CTD were included. The incidence of IM was 0.5% in patients with CTD (32/6911) and the highest in patients with ANCA-associated vasculitis (AAV) (7/480, 1.5%). Molds were isolated in 20 sputum specimens (20/29, 69.0%). *Aspergillus* spp. (81.3%) were the leading strain. Positivity of serum G-test and GM-test was 47.8% (11/23) and 34.6% (9/26), respectively. GM-test was positive in BALF from seven patients. Lung was commonly involved (30/32, 93.8%). Pulmonary nodules (46.7%) and cavitary lesions (36.7%) were common. Ten patients died (31.3%), including three with AAV (42.9%) and seven with SLE (36.8%). Multivariate logistic regression analysis showed that lymphopenia [odds ratio (OR) =3.28, 95% confidential interval (CI) 1.29-8.38, *P*=0.01] and median-to-high dose of glucocorticoid (GC) [OR=3.40, 95% CI 1.04-11.13, *P*=0.04] was associated with IM in patients with CTD. Patients with lymphopenia experienced higher risk of co-infection (50.0% *vs* 0%, *P*=0.01) and mortality (45.5% *vs* 0%, *P*=0.01) compared with patients with normal lymphocyte count.
Conclusion: IM tended to develop in patients with AAV, resulting in high mortality. Sputum culture could be an effective and non-invasive method to diagnose IM. Lymphopenia, and median-to-high dose of GC are associated with IM in patients with CTD.
Acknowledgement: no
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2019-eular.982

Vasculitis

AB0573 **ASSESSMENT OF PRESENCE, SEVERITY AND RISK FACTORS OF POST- THROMBOTIC SYNDROME IN VASCULAR BEHÇET DISEASE: MUTICENTERED RETROSPECTIVE STUDY**

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Background: DVT(deep venous thrombosis) is the most common form of vascular Behçet Disease(VBD).Post-trombotic syndrome(PTS) developing after a thrombotic event in lower extremity is the most important complication of DVT and affects negatively patients' quality of life.
Objectives: We aimed to assess presence,severity and risk factors of PtS and venous disease spesific quality of life in VBD

		Post-thrombotic Syndrome		P Value
		No	Yes	
BSAS score (n:93)		10 (0-47)	20 (1-69)	.002
Standing time (hours)		8.09±3.9	8.24±3.97	.856
VCSS score (n:85)		2.27±1.64	6.54±4.09	.000
VDS	0	18 (50%)	11(19.3%)	.003
	1	15(41.7%)	34(59.6%)	
	2	3 (8.3%)	12(21.1%)	
Veines QoL total score (n:93)		97.19±17.8	77.75±17.3	.000
Veines symptom score		44.3±9.9	35.2±9.1	.000
CEAP		1.53±1.34	2.94±1.83	.000
DVT	1	24(40%)	36(60%)	.888
	2 and over	12 (36.7%)	21 (63.3%)	
Treatment	IS	13 (39.4%)	20 (60.6%)	.817
	IS+AC	17 (36.2%)	30 (63.8%)	
Treatment duration (months)	IS	39(1-200)	36 (3-256)	.600
	AK	12(1-60)	12 (1-156)	.772
Duration of trombosis (years)	<5	15 (40.5%)	22 (59.5%)	.872
	>5	20 (36.4%)	35 (63.6%)	
Compression stocking treatment	No	18 (42.5%)	24 (57.5)	.357
	Yes	12 (24.5%)	27(75.5)	

Methods: This study included 96 BD patients (Female/Male:18/78,mean age: 38.8±8.74)with DVT from 3 tertiary Rheumatology centers in Turkey. When vascular involvement developed, mean age was 32.7±8.65(female: 35.4±10.7; male: 32.09±8; p>.005) Villalta scale is used to assess PTS and according to scale; PTS is present if score >4 and degree of PTS mild, moderate and severe if score 5-9,10-14,>14 respectively. The Venous Disability Score(VDS) and the Venous Clinical Severity Score

(VCSS) were used for the assessment of venous disease. Venous disease-specific QoL was measured through Venous Insufficiency Epidemiological and Economic Study Quality of Life/Symptom (VEINES-QoL/Sym) questionnaire.All patients were reanalyzed using color Doppler USG in the Radiology Department,by a radiologist.In each patient, a total of 16 superficial and deep veins in both legs were assessed for the presence or absence of obstruction, recanalization,reflux, and collaterals within 1 weeks following the clinical examination.

Results: During venous assessment,median disease duration was 9(0-34) years. Eighty(84.2%) patients were under ımmunsupressive(IS) treatment and 13 of these patients were under anticoagulation treatment in addition to ISs. Duration between first vascular event and venous assessment was 6(1-26) years. PTS was present in 57(61.3%) out of 93 patients and severe PTS was present in 19(19.8%)patients.There was no association between the presence of PTS and sex,disease diagnosis age,age during DVT,presence of relaps. There was no difference between patients with or without PTS according to the anticoagulant usage (p=0.817).Doppler ultrasound examination showed bilateral at 31(31.4%) patients and both upper and lower involvment at 40(47.6%) patients.But there is no statically significant relationship between presence of PTS and Doppler findings. In addition to these, there is no statically significant association between PTS and presence of refluxtrombosis at any vessel in the affected leg, but there is a correlation between severe PTS and reflux(r=0.224, p:0.096).VCSS have positive correlation with the presence of reflux(p:0.041, r:0.224). VEINES-QoL/Sym,VCSS and BSAS were significantly worse in patients with PTS.(Table1)

Conclusion: In this study, we found that PTS in lower extremity develops in more than half of the patients with VBD during follow-up, and didn't found any predictor factor for develeopment of PTS. About one third of patient with PTS had severe PTS. PTS is an important clinical problem for physicians treating VBD in daily practice.It should be taken into account as much as preventing vascular relapses during follow-up of patients with VBD.

Table 1. Clinical and characteristic features of PTS

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2019-eular.4890

AB0574

A MONOGENIC DISEASE WITH WIDE RANGE OF SYMPTOMS: DEFICIENCY OF ADENOSINE DEAMINASE 2

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Background: Deficiency of adenosine deaminase 2 (DADA2) is an autosomal recessive autoinflammatory disorder caused by *ADA2* mutations.

Objectives: We aimed to investigate the characteristics of DADA2 patients along with the ADA2 enzyme levels.

Methods: 24 DADA2 patients who admitted to the Adult and Pediatric Rheumatology, Pediatric Haematology, and Pediatric Immunology Departments were included. All exons of the *ADA2* gene were screened by Sanger sequencing in all DADA2 patients. Serum ADA2 enzyme activity was measured by modified spectrophotometric method.

Results: 24 DADA2 patients were included; Group 1, 14 DADA2 patients with polyarteritis nodosa (PAN)-like phenotype; Group 2, 9 patients with Diamond-Blackfan anemia (DBA)-like features and one with immune deficiency. 14 PAN-like DADA2 patients did not have the typical thrombocytosis seen in classical PAN. Inflammatory attacks were evident in only Group 1 patients. Serum ADA2 was low in all DADA2 patients except one who was tested after hematopoietic stem cell transplantation. There was no significant difference in ADA2 levels between PAN-like and DBA-like DADA2 patients (Figure 1). ADA2 activities of heterozygote family members were about half the level of the control subjects. However, in heterozygote DADA2 patients, serum ADA2 levels were as low as the ones of homozygote DADA2 patients. *ADA2* mutations were affecting the dimerization domain in Group 1 patients and in the catalytic domain in Group 2 patients (Table 1).

Table 1. Molecular results of *ADA2* gene analyses in DADA2 patients (D, dimerization; C, catalytic)

Group number	Patient numbers	Mutation position	Mutation type	The affected domain of ADA2 protein
1	11	Exon 2: c.139G>A	Homozygous missense	D
1	1	Exon 2: c.139G>A/ Exon 2: c.140G>T	Compound heterozygous missense	D
1	2	Exon 2 c.139G>A	Heterozygous missense	D
2	1	Exon 4: c.620T>C/ Exon 9: c.1360G>C	Compound heterozygous missense	C
2	2	Exon 7: c.1072G>A	Homozygous missense	C
2	1	Exon 4: c.629delT/ Exon 2: c.144_145ins	Compound heterozygous Del/ins_frameshift and non-sense	C
2	1	Exon 10: c.1445A>G	Homozygous missense	C
2	1	Exon 4: c.680-681delAT	Homozygous deletion-Frameshift non-sense	C
2	1	Exon 9: c.1367A>G	Homozygous missense	C
2	2	Exon 6: c.916C>T	Homozygous non-sense	C
2	1	Exon 9: c.1392_1393insG	Homozygous missense	C

Conclusion: We suggest that enzyme activity of ADA2 should be assessed along with genetic analysis since there are heterozygote patients with absent enzyme activity. Our data confirms a possible genotype phenotype correlation where dimerization domain mutations are associated with a PAN-like phenotype whereas catalytic domain mutations are associated with hematological manifestations.

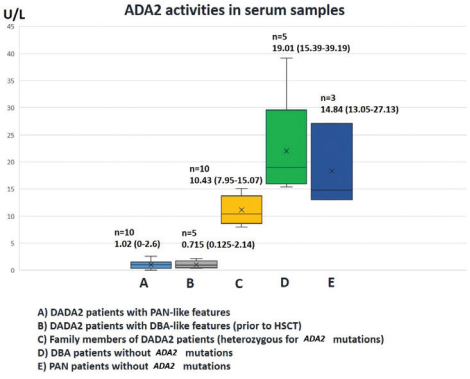

Figure 1

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Acknowledgement: This study was supported by Scientific and Technological Research Council of Turkey (TÜBITAK) with grant numbers of 315S192. Prof. Nurten Akarsu performed the molecular studies of DBA patients and special thanks to her for great support.

Disclosure of Interests: Ezgi Deniz Batu: None declared, Ekim Z. Taskiran: None declared, Hatice Asuman Özkara: None declared, Şule Ünal: None declared, Naz Guleray: None declared, Abdulsamet Erden: None declared, Omer Karadag: None declared, Fatma Gümrük: None declared, Mualla Çetin: None declared, Yelda Bilginer: None declared, Hafize Emine Sonmez: None declared, Deniz Cagdas Ayvaz: None declared, Ilhan Tezcan: None declared, Seza Özen Consultant for: Seza Ozen is receiving consultancy fees from Novartis, Speakers bureau: Roche
DOI: 10.1136/annrheumdis-2019-eular.584