THU0574

DOES THE USE OF COLCHICINE EFFECT COGNITIVE FUNCTIONS IN FAMILIAL MEDITERRANEAN FEVER? PRELIMINARY STUDY

Halise Hande Gezer^{1,1}, Ozge Devezer Uslu², Didem Erdem¹, Sevtap Acer Kasman¹, Mehmet Tuncay Duruöz¹. ¹Marmara University School of Medicine, Department of Physical Medicine and Rehabilitation, Rheumatology Division, Istanbul, Turkey; ²Institution of Clinical and Forensic Psychology, Clinical Psychology, Istanbul, Turkey

Background: In Familial Mediterranean Fever (FMF), although cognitive functions have been shown to be impaired in children and adolescents, it has been shown that colchicine can be preservative on the cognitive functions in patients who are on long term colchicine treatment.

Objectives: This study aimed to evaluate cognitive functions in adult patients with FMF and cognitive effects of colchicine use.

Methods: The study included patients who were diagnosed with FMF according to Tel-Hashomer criteria. The control group included patients with no other inflammatory or systemic disease. Clinical features such as disease duration, comorbid diseases, colchicine treatment duration and dosage, amyloidosis and chronic renal failure (CRF), FMF gene mutation and PRAS scoring for evaluation of disease activity were recorded. Pittsburgh Sleep Quality Index (PSQI), Fatigue Severity Scale, FMF Quality of Life Scale and Beck Depression Scale were used to assess patients clinical situations. Cognitive measurements were evaluated under executive-propellent functions. KAS test was used for information processing and fluency skills; fruit-name test was used for the evaluation of focusing, concentration, and attention skills; animal counting test and The Montreal Cognitive Rating Scale (MOCA) which can evaluate different subunits (visuospatial/executive, naming, memory, attention, language, abstraction, delayed recall, and orientation) were used for the fluency and maintenance of attention. Different attention parameters were evaluated for patients and healthy individuals. These attention parameters were; focusing, elaborate, sustainability, ability to pay attention to two information at the same time.

Descriptive analysis was performed for all parameters. Mann-Whitney U-test and Spearman correlation coefficient were used to compare parameters. $P < 0.05$ was accepted as statistically significant.

Results: The study included 24 (21 women, 3 men) patients with FMF and 10 (7 female, 3 male) age, sex and BMI matched healthy controls. The mean age of the patients and controls were 36.83 (SD:10.9) and 39.3(SD:8.6), respectively. No significant difference was found between FMF and healthy control groups regarding animal counting, KAS test, MOCA test, and subgroups. Only fruit-name counting test was decreased in the FMF group compared to the healthy controls ($p < 0.05$). As the duration of colchicine treatment was prolonged, a moderate positive correlation was found in KAS scores ($r=0.511$) and MOCA naming scores ($r=0.445$). In the FMF group, the number of attacks in the last three months and the sleep scores of Pittsburgh and depression scores had a moderate positively correlation ($r = 0.496$). Depression scores and quality of life scores were highly correlated ($r = 0.631$).

Conclusion: FMF patients attention parameters are impaired compared to the healthy controls. Information processing and fluency performance is increased in FMF patients with the duration of colchicine treatment that demonstrate the ability to categorize and fluent use of the information. Number of attacks are correlated with poor sleep quality and depression.

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THU0575

HYPERCOAGULABILITY AS A CAUSE OF THROMBOSIS IN BEHÇET'S SYNDROME: A SYSTEMATIC REVIEW AND META ANALYSIS

Gul Guzelant Ozkose, Berna Yurttas, Sinem Nihal Esatoglu, Vedat Hamuryudan, Hasan Yazici, Gulen Hatemi. *Istanbul University-Cerrahpasa, Medical Faculty of Cerrahpasa, Internal Medicine, Division of Rheumatology, Istanbul, Turkey*

Background: While thrombosis in Behçet's Syndrome (BS) is considered to be mainly caused by inflammation in the vessel wall, several pro-thrombotic factors have been studied with inconsistent results.

Objectives: We aimed to perform a systematic review of clinical studies investigating the thrombophilic factors in BS.

Methods: The online database of PubMed was searched with the key-word "Behcet*" in four languages (English, German, French and Turkish) from inception up to May 2018. Titles and/or abstracts of all studies were screened independently by two reviewers (GG and BY) for studies reporting on thrombosis, fibrinolysis, endothelial factors and comparing BS patients with and without thrombosis. Conflicts were solved by a third reviewer (GH). The pooled odds ratios (OR) with 95%CI were calculated for binary outcomes and standardized mean differences (MD) were calculated for continuous outcomes by using RevMan 5.3.

Results: Of 9937 articles, 9373 were excluded due to repetition and inappropriate study design after reviewing titles and abstracts. Full text review of the remaining 564 articles yielded 86 papers meeting our predetermined inclusion criteria.

Several factors such as protein C, protein S, active protein C resistance, anti-thrombin III, plasminogen, plasminogen activator inhibitor, fibrinogen, factor 7, factor 12, thrombin activatable fibrinolysis inhibitor, antidiolipin antibodies, antiβ2 Glycoprotein1 antibodies and methylenetetrahydrofolate reductase gene C677T mutation were not different in BS patients with thrombosis compared to those without thrombosis. On the other hand, vascular endothelial growth factor levels, P-selectin glycoprotein ligand-1, platelet-activating factor seemed to be more frequent in BS patients with thrombosis in the few studies reporting on these, including a small number of patients.

Among the 11 parameters with controversial results across studies, meta-analysis showed significantly higher homocysteine levels, higher factor 8 levels, more frequent Factor V Leiden mutations and higher von Willebrand factor levels in BS patients with thrombosis, whereas the pooled difference was not significant for mean platelet volume, tissue plasminogen activator, prothrombin gene mutations, lupus anticoagulant, P-selectin level, erythrocyte aggregation and thrombomodulin level (Table).

Conclusion: Among the several prothrombotic factors that were studied in BS patients, factor V Leiden mutation, high homocysteine levels, factor 8 levels and von Willebrand factor levels may be associated with thrombosis in BS. Studies investigating these factors together in a large number of patients together with appropriate controls are needed to confirm these results.

Table. Meta-analysis of studies with controversial results

Prothrombotic Factor	Number of studies	Number of Behçet's patients		MD/OR (95% CI)
		With thrombosis	Without thrombosis	
vWF (U/dl)	3	37	93	MD: 10.09 (2.70, 17.47)
Factor V Leiden	9	204	446	OR: 2.17 (1.32, 3.57)
Homocysteine	12	285	436	MD: 1.26 (0.86, 1.66)
FVIII level	3	53	128	MD: 14.68 (2.87, 26.49)
tPA	5	103	200	MD: 0.09 (-0.47, 0.66)
Prothrombin mutation	7	189	398	OR: 1.63 (0.85, 3.13)
Mean platelet volume	5	73	327	MD: 0.13 (-0.13, 0.39)

Lupus anticoagulant	3	37	85	OR: 3.66 (0.98, 13.72)
P-selectin	2	33	75	MD: 0.85 (-0.08, 1.77)
Thrombomodulin	5	92	264	MD: -0.22 (-0.86, 0.41)
Erythrocyte aggregation	3	47	112	MD: -0.24 (-0.62, 0.15)

MD: mean difference, OR: odds ratio, tPA: tissue plasminogen activator

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THU0576 PROGNOSTIC FACTORS PREDICTING THE SURVIVAL OF PATIENTS WITH MACROPHAGE ACTIVATION SYNDROME

Seungmin Jung, Sungsoo Ahn, Sangwon Lee, Jason Jungsik Song, Yongbeom Park. *Yonsei University College of Medicine, Internal Medicine, Seoul, Korea, Rep. of (South Korea)*

Background: Macrophage activation syndrome (MAS) is a hyperinflammatory condition, which can lead to death in patients with rheumatologic disease. However, the prognosis of adult patients with MAS is largely unknown.

Objectives: We aimed to investigate the mortality in patients with MAS accompanied Adult-onset Still's disease (AOSD) or systemic lupus erythematosus (SLE) and to evaluate the prognostic factors predicting the mortality in patients with MAS.

Methods: We retrospectively reviewed febrile patients with AOSD or SLE admitted to Severance Hospital between 2005 and 2018. Patients who satisfied the classification criteria of MAS was included in the analysis. Cox-regression analysis was performed to evaluate the clinical factors associated with the overall mortality in patients with MAS.

Results: Of the total 123 patients, 48 (39%) patients and 75 (61%) patients were diagnosed with AOSD and SLE, respectively. Forty-three patients (35%) were died from MAS during hospitalization. There was no significant difference in mortality between AOSD and SLE ($P = 0.675$). In multivariate analysis, cytopenia, and insufficient reduction of ferritin and rebound of inflammation after initial treatment with glucocorticoid were independently associated with death in patients with MAS ($P = 0.019$, < 0.001 , and 0.011 , respectively).

Conclusion: The presence of cytopenia, and treatment response after glucocorticoid treatment was closely related with death in patients with MAS. More intensive treatment should be considered in high-risk patients with poor prognostic factors.

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THU0577 EFFICACY OF RADIOSYNOVIORTHESIS IN PIGMENTED VILLONODULAR SYNOVITIS OF THE KNEE

Zsolt Karoly Mangel, György Nagy, Margit Szentesi. *Polyclinic of the Hospitaller Brothers of St. John of God, Rheumatology, Budapest, Hungary*

Background: Pigmented villonodular synovitis (PVNS) is a rare disorder with the benign tumoral proliferation of the synovium. The surgical treatment of PVNS alone in most cases is unsatisfactory, because if a few cells have not been removed, the disease will recur.¹ Post-synovectomy adjuvant treatment with intra-articular injection of Yttrium-90 (⁹⁰Y) or Holmium-166 (¹⁶⁶Ho) yielded better results. The radiosynoviorthesis (RSO) is an effective way of treating the chronic synovitis, with this method we are eliminate the inflammation in 75 percent of the cases.²

Objectives: To study efficacy of radiosynoviorthesis after the surgical synovectomy in pigmented villonodular synovitis of the knee.

Methods: Between May 1986 and August 2018, 17 patients (seven men and ten women aged 14–68 years) with diffuse PVNS were treated. All patients had monoarticular arthritis of the knee with histologically proved PVNS. The patients underwent 33 operations, two patients had four surgical procedures, one patient underwent three surgeries, eight patients had two surgeries and six patients had one surgical procedure (Table 1.). The radiosynoviorthesis was performed according to the method accepted in the national protocol. Yttrium-citrate injectable suspension marked by 185 MBq ⁹⁰Y-citrate injectable suspension, and 40 mg of 1 ml triamcinolone acetonide and 1 ml of lidocaine 1%.³ Holmium-phytate injectable suspension marked by 600 MBq ¹⁶⁶Ho-phytate injectable suspension, and 40 mg of 1 ml triamcinolone acetonide and 1 ml of lidocaine 1%.³

Evaluation was based on the criteria as described by Müller, Rau and Schütte the score system was developed by the authors.⁴ The circumference of joint-swelling, the joint function, the measure of flexio-contracture, pain in state of rest and load on a pain analogue scale 1-10, joint warmth, walking capacity, the numbers of joint-punctures after the treatment, whether operation was necessary or not after the treatment were examined.

Number of patients	age	surgery before RSO				number of RSO	
7 men and 10 women total: 17	14-68 years	1	2	3	4	1	2
6 patients	8 patients	1 patient	2 patients	10 patients	6 patients	1 patient	

Table 1. Patients with pigmented villonodular synovitis

Results: Mean follow-up time was 56 months (range from 4 to 144 months). All patients were followed up using clinical assesment. After the first injection excellent and good results were recorded in 41%. After the second radiosynoviorthesis thirteen patients (76%) showed excellent and good response (Table 2.). The mean functional evaluation score of 17 patients was 28 (range 16-34). Most of the ratings were excellent or good, in four cases moderate (24%). No complications were noticed after surgery or after the radiosynoviorthesis.

functional evaluation score	response	patients	%
29-35	excellent	10	59%
22-28	good	3	18%
15-21	moderate	4	23%
8-14	mild	0	
0-7	worsening	0	

Table 2. Functional evaluation score after the RSO

Conclusion: A combination of debulking surgery with radiosynoviorthesis of Yttrium or Holmium for diffuse PVNS of the knee joint is a reliable treatment method, with good results.

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