Figure:

	Gender	Age at onset of initial disease (median, range)	Presenting symptoms	Bilateral	Laboratory Markers, (median)	Extracranial muscle involvement	Optic nerve involvement	Localized gland involvement	Referred involvement
Idiopathic inflammatory pseudotumor (n=16)	F/M:1/15	35 (19-71)	Swelling (n=6), Pain (n=4), Erythema (n=7), Vision loss (n=2), Diaphous (n=2)	n=5 (31.2%)	ESR: 18 (2-60) CRP: 0.4 (0.1-4.6)	11 (68.7%) LDM (n=10), MDM (n=1) SDM (n=1), SOM (n=1)	4 (25.0%)	5 (31.2%)	2 (12.5%)
SpA-related disease (n=13)	F/M:1/12	40 (21-59)	Swelling (n=7), Pain (n=8), Purpura (n=6), Pneumonia (n=1), Vision loss (n=1), Blurred Vision (n=1)	n=3 (23.1%)	ESR: 14 (2-29) mm/h CRP: 0.4 (0.1-5.4) mg/dL dIgM level: 420 (122-3790)	7 (53.8%) LDM (n=4), MDM (n=3) SDM (n=1), SOM (n=2) IDM (n=1), SOM (n=2)	4 (30.8%)	7 (53.8%)	3 (23.1%)
GPA (n=7)	F/M:3/4	44 (5-64)	Swelling (n=2), Pain (n=2), Vision loss (n=1), Diaphous (n=1)	n=3 (42.9%)	ESR: 41 (7-108) mm/h CRP: 1.5 (0.2-29) mg/dL ANCA (n=6, 85.7% +)	6 (85.7%) LDM (n=4), MDM (n=4) SDM (n=1), SOM (n=1)	4 (57.1%)	6 (85.7%)	1 (14.3%)
Thyroid-associated orbitopathy (n=1)	M	54	Pain: periorbital	Unilateral	ESR Normal ESR: 13 mm/h CRP: 0.3 mg/dL	LDM/MDM/SDM	-	+	+
Sjögren's syndrome (n=1)	M	49	Pain: swelling	Unilateral	ANA (+), ESR Normal ESR: 10 mm/h CRP: 0.3 mg/dL	-	-	+	+
Relapsing polychondritis (n=1)	M	47	Pain: swelling	Unilateral	ESR: 47 mm/h CRP: 10 mg/dL	LDM	+	+	+
Fungal granulomatous angitis (n=1)	F	75	Pain: erythema vision loss	Unilateral	ESR: 17 mm/h CRP: 1.2 mg/dL	SDM/IDM/SDM	+	+	+

GPA: Granulomatosis with polyangiitis; ESR: Erythrocyte sedimentation rate; CRP: c-reactive protein; ANCA: Anti-neutrophil cytoplasmic antibody; ANCA: Anti-neutrophil cytoplasmic antibody; ANCA: Anti-neutrophil cytoplasmic antibody; ANCA: Anti-neutrophil cytoplasmic antibody.

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AB1039

LL-37, IL-36, GALECTIN-3 AND TLR-3 LEVELS IN IDIOPATHIC GRANULOMATOUS MASTITIS

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Background: Idiopathic granulomatous mastitis (IGM) is a non-infectious inflammatory disorder of the breast characterized by non-caseous granulomas. It is a chronic granulomatous inflammatory disease frequently seen in young fertile women, the cause of which has not been clearly understood. Immunosuppressive agents and surgical interventions are used in the treatment.

LL-37 is a cathelicidin-derived antimicrobial peptide with immunomodulatory properties that are effective in innate immunity. In addition, IL-36, galectin-3, TLR-3 are effective in autoimmunity with proinflammatory properties.

Objectives: With this study, we aimed to investigate the potential alterations of LL-37, IL 36, Galectin 3 and TLR-3 levels in IGM.

Methods: 35 female patients with biopsy-confirmed IGM and 35 healthy controls were included in the study. The serum samples of the subjects LL-37, IL 36, Galectin 3 and TLR-3 levels were studied using the Elisa method. While studying LL 37 and Galectin 3 levels in the tissue, samples of 10 patients who underwent mastoplastic for cosmetic reasons were used for the control group. Ten patients whose paraffin blocks were eligible for re-study were included in the study for tissue examinations. Based on the prevalence (0.1: <25%, 0.4: 26-50%, 0.6: 51-75%, 0.9: 76-100) and

severity (0: no, +0.5: very little, +1: little, +2: medium, +3: severe) of immunoreactivity in staining, histoscore was created (histoscores = prevalence x severity). The data were evaluated using appropriate statistical analysis and p <0.05 was considered statistically significant.

Results: When the patient and control groups included in the study were compared, there was no significant difference in age. In serum samples, LL 37, IL 36, Galectin 3 and TLR 3 levels were statistically significantly lower in IGM group compared to the control group (p <0.001 for each) (Table-1). In biopsy samples, LL 37 level was found to be significantly lower in IGM group compared to the control group (p <0.001). However, no significant difference was detected in Galectin 3 levels in tissue studies (Table-2).

Conclusion: In our study, we found that the levels of LL 37, IL 36, Galectin 3 and TLR 3 decreased in serum samples in IGM disease whose etiology was not clearly understood. In addition, we showed that in patients with IGM, LL 37 levels decreased at the tissue level. Studies have shown that in cases of severe sarcoidosis, LL 37 deficiency is reduced both in level and gene expression. So they thought, deficiency of cathelicidin LL 37 may impede resolution of inflammation in the tissue of patients with severe form disease.

References:

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Table 1. LL 37, IL 36, Galectin 3 and TLR 3 levels in serum samples

	IGM (N:35)	CONTROL (N:35)	P
Age	35,92 ± 5,19	34,03 ± 3,81	0,08
LL 37 (ng/ml)	5,20 ± 5,48	40,05 ± 31,97	<0,001
IL 36 (pg/ml)	294,74 ± 125,94	864,71 ± 442,39	<0,001
Galectin 3 (ng/ml)	3,61 ± 3,16	15,53 ± 10,14	<0,001
TLR 3 (pg/ml)	931,49 ± 443,86	4019,36 ± 2599,51	<0,001

IGM: Idiopathic granulomatous mastitis

Table 2. Evaluation of LL-37 and Galectin 3 levels with histoscores in biopsy samples

	IGM (N:10)	CONTROL (N:10)	P
LL 37	0,006 ± 0,025	0,140 ± 0,516	<0,001
Galectin 3	0,293 ± 0,201	0,400 ± 0,290	0,32

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AB1040

COEXISTENCE OF DEMYELINATION DISEASE AND FAMILIAL MEDITERRANEAN FEVER

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Table 1. Some clinical and laboratory features of patients with FMF or MEFV mutations accompanied by demyelination disease

Cases	Age/Sex	Diseases	MEFV mutations	The onset age/ diagnostic age for FMF	The onset age for DD/MS/ Presenting manifestations/MRI findings	Treatment for FMF / DD/MS
Case 1 (F1)	17/F	FMF+DD	M694V homozygous	3/5	15 OB (-) Fusiform plaques in the cingulate gyrus; plaques in T4-6	Colchicine IL-1 RA Glatiramer acetate
Case 2 (F1)	46/F	FMF+MS	M694V homozygous	8/9	28 Optic nerve involvement OB(+) Plaques (+)	Colchicine Glatiramer acetate
Case 3 (F1)	17/F	FMF+MS	M694V heterozygous	3/5	15 Loss of the right eye, vertigo OB(+) Plaques	Colchicine Pulse steroid Beta-interferon Teriflunomide
Case 4 (F2)	36/F	MS+MEFV mutation	M694V/R202Q	-	27 Headache, blurred vision, optic nerve atrophy OB(+) Plaques (+)	Glatiramer acetate
Case 5 (F2)	16/F	MS?+FMF+Cutaneous vasculitis	M694V/R202Q	16/16	11 Headache, blurred vision No LP (denied by pt) Plaques	-

F: Female, F1: Family 1, F2: Family 2; DD: Demyelination disease; MS: Multiple sclerosis; MRI: Magnetic resonance imaging; OB:Oligoclonal band; LP: Lumbar puncture