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AB1304 COMPARISON OF CHARACTERISTICS OF PATIENTS WITH IDIOPATHIC GRANULOMATOSIS MASTITIS IN REMISSION AND NON-REMISSION: RESULTS FROM A TERTIARY REFERRAL CENTRE

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Background: Idiopathic granulomatous mastitis (IGM) is an uncommon, benign, chronic inflammatory disease of the breast of unknown etiology. Although an ideal treatment regimen is controversial in IGM, the ultimate goal is remission.
Objectives: To compare the clinical, radiological, laboratory and treatment features of patients in remission and non-remission, and assess predictor factors of remission.
Methods: The data of patients who were histopathological diagnosed with IGM between 2010 and 2020 were evaluated retrospectively. The patients were divided into 2 groups as those in complete remission and non-remission. Complete improvement in clinical and physical examination with medical and/or surgical intervention findings at 2 consecutive visits was defined as complete remission. Patients who were not in complete remission including partial remission and refractory to treatment, were defined as non-remission group.
Results: In this study, there were a total of 103 patients with a diagnosis of IGM followed-up in the rheumatology clinic. Of these, 39 (38%) were in remission and 64 (62%) were non-remission. Age, age at symptom onset, comorbidity, pregnancy (ever), disease localization, clinical signs and symptoms, imaging classification, and baseline acute phase reactants and autoimmune markers were similar in the remission and non-remission groups. The use of antibiotics and azathioprine was significantly higher in the remission group than in the non-remission group. However, there was no difference between the groups in terms of breast surgery, use of corticosteroids and methotrexate (Table 1). In the univariable analysis, antibiotic use [OR: 2.55 (95% CI 1.07-6.11)] was found to be a predictor factor of remission.
Conclusion: Patients with in remission did not have the distinctive baseline demographic, clinical, laboratory and radiological features that distinguish them from patients with in non-remission. Patients in remission used more antibiotics and azathioprine. Antibiotic use seems to be associated with remission in univariable analysis. However, further studies are needed to evaluate the factors associated with remission in IGM.
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AB1305 A SYSTEMATIC REVIEW OF AA AMYLOIDOSIS AMONG PATIENTS WITH BEHÇET'S SYNDROME

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Background: Data on patients with Behçet's syndrome (BS) complicated with AA amyloidosis is limited to case reports or case series with a small number of patients.
Objectives: In this study, we aimed to perform a systematic review (SR) of published reports on BS patients with AA amyloidosis.
Methods: PubMed and EMBASE were searched with the keywords "Behcet* AND amyloidosis", without date and language restriction, until May 2020. Two independent reviewers (SNE, GK) performed title/abstract and full text screening and data extraction. A third reviewer (GH) made the final decision in case of disagreement between the two reviewers. Studies that reported patients who were reported by authors as having BS and AA amyloidosis were included. The risk of bias assessment was done using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) tool.
Results: The systematic literature search yielded 760 articles of which 703 were excluded after title and abstract review. After full-text review, we further excluded 15 duplicate articles and 1 article was added after handsearching the reference lists of the full texts. Finally, we included 43 articles reporting 96 cases. Among these articles, 38 were case reports and 5 were case series reporting between 6 and 14 patients. All patients but 8 were reported from Mediterranean countries.

Table 1. Characteristics of patients in remission and non-remission

	All patients	Patients in remission	Patients not in remission	p value
	n= 103	n = 39	n= 64	
Age, years	38.1 ± 7.1	39.6 ± 7.8	37.2 ± 6.5	0.07
Age at symptom onset, years	33.5 ± 6.6	34.4 ± 7.3	33 ± 6.1	0.30
Comorbidity, n (%)				
Thyroid disorders	4 (3.9)	1 (2.6)	3 (4.7)	1.00
Hypertension	2 (2)	2 (5)	0	0.14
Diabetes mellitus	1 (1)	1 (2.6)	0	0.38
Pregnancy (ever), positive/total (%)	75/77 (97.4)	31/32 (97)	44/45 (98)	1.00
Breast localization, n (%)				
Left	47 (45.6)	19 (48.7)	28 (43.8)	0.88
Right	36 (35)	13 (33.3)	23 (35.9)	
Bilateral	20 (19.4)	7 (17.9)	13 (20.3)	
Clinical signs and symptoms, n (%)				
Palpabl mass	94 (91.3)	35 (89.7)	59 (92.2)	0.72
Fistula formation	37 (38.1)	16 (42.1)	21 (35.6)	0.52
Erythema	61 (65.6)	23 (65.7)	38 (65.5)	0.98
Nipple inversion	11 (12)	2 (5.4)	9 (16.4)	0.19
Axillary lymph node	45 (48)	17 (47.2)	28 (48.3)	1.00
Erythema nodosum	10 (9.9)	4 (10.3)	6 (9.7)	1.00
Arthralgia	13 (12.6)	3 (7.7)	10 (15.6)	0.36
BI-RADS classification*, n (%)				
2	13 (25)	7 (35)	6 (18.8)	0.12
3	31 (59.6)	9 (45)	22 (68.8)	
4	6 (11.5)	2 (10)	4 (12.5)	
5	2 (3.8)	2 (10)	0	
Baseline laboratory				
-CRP (mg/dL)	0.75 (1.4)	0.75 (1.4)	0.73 (1.4)	0.77
- ESR (mm/h)	23 (28)	21.5 (28)	25.5 (28)	0.39
-ANA (more than or equal 1/100), n$^{\alpha}$ (%)	37/69 (53.6)	12/25 (48)	25/44 (56.8)	0.48
-RF, n$^{\alpha}$ (%)	12/30 (40)	5 /16 (45.5)	7/19 (36.8)	0.71
-anti-CCP, n$^{\alpha}$ (%)	3/12 (25)	2/5 (40)	1/7 (14)	0.52
Treatment, n (%)				
Surgical intervention	38 (36.9)	16 (41)	22 (34.4)	0.49
Antibiotics	63 (61.2)	29 (74.4)	34 (53.1)	0.03
Corticosteroids	93 (90.3)	34 (87.2)	59 (92.2)	0.49
Methotrexate	66 (64.1)	26 (66.7)	40 (62.5)	0.67
Azathioprine	19 (18.4)	11 (28.2)	8 (12.5)	0.04

* 52 patients with data were evaluated.$^{\alpha}$ positive/total
Results are given as mean ± SD or median (IQR).
ANA: Antinuclear antibody; anti-CCP: Anti-cyclic citrullinated peptides; BI-RADS: Breast imaging-reporting and data system; RF: Rheumatoid factor

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The quality of all articles according to GRADE was very low due to the lack of a control group.
The main features of the patients were male predominance (81/96, 84%), a high frequency of major organ involvement (62/80, 77.5%) especially vascular involvement (60%), a low frequency of comorbidities predisposing to AA amyloidosis (11/96, 11.5%), and a very low frequency of gastrointestinal involvement (3/72, 4%). All but 8 patients were diagnosed with BS and AA amyloidosis simultaneously. The most common presentation was nephrotic syndrome (60/81, 74%). Presenting symptoms other than proteinuria were diarrhea (n=2), acute renal failure (n=2), upper gastrointestinal bleeding (n=1), end stage renal disease (ESRD) (n=1), cardiac symptoms due to cor pulmonale (n=1), and hypertension (n=1). Renal biopsy (72%) and rectal biopsy (17%) were the most commonly used procedures to diagnose AA amyloidosis.
After diagnosing AA amyloidosis, colchicine was initiated in 58 patients, cyclophosphamide in 16, and biologics in 3 (1 anakinra and 2 tocilizumab). In the 67 patients with available data on follow-up, 43% of the patients were followed-up for ≤1 year and median follow-up duration was 20 months (IQR: 4-48). Among the 64 patients with available data, 30 (47%) had developed ESRD. Among the 72 patients with available data on survival status, 30 patients (42%) had died. Ten patients (33%) had died within 6 months, 15 had died after a median follow-up of 48 months (IQR: 24-150), and follow-up duration was not available in the remaining 5 patients including 3 patients whose diagnoses were made by autopsy. Reasons for death were infection (n=7), ESRD (n=6), intractable diarrhea (n=3), pulmonary embolism (n=1), cor pulmonale (n=1), hemorrhage due to pulmonary artery aneurysm (n=1), liver cirrhosis (n=1), gastric cancer (n=1), subarachnoid hemorrhage (n=1), and not reported (n=8).
Conclusion: Male gender and major organ involvement, especially vascular involvement, appear to be risk factors for the development of AA amyloidosis in BS patients. While BS patients complicated with AA amyloidosis have been reported rarely, it is a fatal complication of BS. One third of the patients had died within 6 months after AA amyloidosis diagnosis.
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AB1306 AA AMYLOIDOSIS IN CHRONIC RHEUMATIC DISEASES: ABOUT 20 CASES

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Background: Secondary AA amyloidosis is a complication of certain uncontrolled chronic inflammatory rheumatic diseases. The diagnosis of AA amyloidosis is anatomo-pathological based on the detection of amyloid deposits within a biopsy specimen.
Objectives: The aim of this study is to determine the clinical, biological, therapeutic and evolutionary aspects of amyloidosis during these diseases.
Methods: This is a retrospective descriptive study conducted in the rheumatology department in Monastir, Tunisia, including patients followed for chronic inflammatory rheumatism complicated by AA amyloidosis.
Results: Our study included 20 patients (13 women and 7 men). Their mean age at the time of diagnosis of amyloidosis was 55.4 years [18-76]. The rheumatic diseases were: Rheumatoid Arthritis (RA) in 13 cases (65%), Ankylosing Spondylitis in 3 cases (15 %), Behçet's disease in 2 cases and Juvenile idiopathic arthritis in its systemic form in 2 cases (10% for both). Amyloidosis occurred after a mean duration of rheumatic disease of 11.5 years [3-27]. The mean sedimentation rate at diagnosis of amyloidosis was 100.5 mm per hour [21-131]. The mean C-reactive protein at diagnosis was 38.35 mg/L [1.4-135]. Inaugural manifestations of amyloidosis were: edema of lower limbs in 8 cases, nephrotic syndrome in 7 cases, renal failure in 4 cases and an abdominal clinical presentation (diarrhea and abdominal pain) associated with proteinuria in 2 cases. Proteinuria was constant, with a mean level of 7.5 g/24 h [3.5-20]. It was isolated in 5 cases. Amyloidosis was subclinical in 2 cases and was discovered by an abdominal fat biopsy performed during a research protocol. The diagnosis was confirmed by: renal biopsy (n=9), rectal biopsy (n=5), salivary gland biopsy (n=4) and abdominal fat biopsy (n=2). Amyloidosis extension was assessed for 50% of our patients. Cardiac involvement was the most frequent, revealed in 5 patients. Concentric left ventricular hypertrophy was found in 4 patients. Pulmonary arterial hypertension and pericardial effusion were found in 1 case. Hepatic involvement was detected in 2 cases in the form of hepatomegaly without disturbance of liver function tests. Adrenal gland involvement was detected in 1 case with homogeneous hypertrophy of the adrenal glands associated with adrenal dysfunction. Only 1 patient had peripheral nerve damage in the form of bilateral carpal tunnel syndrome. Concerning the treatment; 12 patients received Colchicine. Chloraminophen was tried for 4 patients with amyloidosis secondary to RA and Azathioprine for one patient with RA. The evolution was unfavorable in 6 cases marked by death within a mean time of 3.4 years.
Conclusion: Although its prevalence is decreasing, owing to therapeutic progress in chronic rheumatic diseases, AA amyloidosis remains a not exceptional complication. It has a poor prognosis, particularly because of renal disease and cardiac involvement.
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AB1307 RHEUMATIC SYNDROMES INDUCED BY ONCOLOGIC AND HEMATOLOGIC TARGETED THERAPIES

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Background: Targeted therapy has gradually become the first-line clinical therapy for solid tumors and hematologic disorders (1). Immune related adverse side effects (irAE) due to checkpoint inhibitors are well recognized side effects in patients with underlying oncologic disorders; however, information regarding rheumatic side effects due to several targeted therapies are rarely reported.
Objectives: Our aim was to describe the main rheumatic syndromes induced by targeted therapies.
Methods: This observational study included patients from 2 referral University centers in Barcelona. We included patients referred from Oncology and Hematology departments visited from January 2017 to January 2022 at Hospital Clinic and Hospital de Bellvitge, Barcelona, Spain.
Results: Fifteen patients were included (Female 60%), with a mean age 63.3± 10.8 years. Nine patients had hematological disorders and 8 solid tumors. Main diagnosis included Multiple Myeloma in 3 (20%) patients, Non-Hodgkin Lymphoma 2 (13%), Acute Myeloid Leukemia 2 (13%), Melanoma 2 (13%), Colon carcinoma 1 (6.7%), Breast carcinoma 1 (6.7%), Glioblastoma multiforme 1 (6.7%), Chronic lymphatic leukemia 1 (6.7%), Renal cell carcinoma 1 (6.7%) and Sésary Syndrome in 1 (6.7%) patient. Targeted therapies included MAP kinases in 4 (26.7%) patients, Anti CD-38 in 3 (20%), Anti BRAF in 2 (13.3%), and anti STAT-3, anti VEGF, BTK inhibitor, anti CCR4, Isocitrate dehydrogenase 2 inhibitor and Halichondrin B analoge in 1 (6.7%) patient each. Treatment was administrated as monotherapy in 9 (60%) and as combined therapy in 6 (40%). Mean time from the initiation of targeted therapies and rheumatic syndrome onset was 5.0 ± 3.8 months. Main rheumatic syndrome were: Polymyalgia rheumatica (PMR) in 4 (26%) patients, Raynaud's phenomenon in 2 (13%), Tenosynovitis in 2 (13%), RA-like 2 (13%), cutaneous vasculitis, amyopathic dermatomyositis, oligoarthritis, spondyloarthropathy-like and non-inflammatory arthralgias in 1 (6.7%) case each. Main clinical characteristics are summarized in Table 1. In 8 (53%) cases, targeted therapies were stopped due side effects. Rheumatic syndrome were treated with, glucocorticoids (GC) 12 (80%), calcium antagonist 2 (13%), Hydroxychloroquine in 2 (13%), Methotrexate in 1 (6.7%), Sulfasalazine in 1 (6.7%) and NSAID in 1 (6.7%) patient. Mean follow-up were 15.4± 20 months. Most of the patients remain alive (86%). According oncologic prognosis, 4 (26%) had complete response, 6 (40%) had partial response and 5 (33%) had disease progression. At the time of the last visit, 6 (40%) patients remain on GC (mean dose 6.7 ± 4.7 mg/d) and 4 (26%) patients on DMARD therapy.
Conclusion: In this heterogeneous group of patients treated with targeted therapies, we described a variety of induced rheumatic syndromes including PMR, arthritis, and even vasculitis or dermatomyositis. An important proportion of patients stopped the treatment due to side effects and some of them required sustained treatment including GC and DMARD.
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