Table 1. Pre- and post-treatment laboratory and clinical findings.

	Pre-treatment	Post-treatment	P value
ESR (mm/h)	42.45±28.88	17.24±11.31	<0.001*
CRP (mg/L)	24.72±35.32	5.37±6.5	0.004*
Mass Size (mm)	36.94±16.40	10.79±15.01	<0.001*
Fistula (n)	15/33 (45%)	2/33 (6%)	<0.001**
Nipple discharge(n)	17/33(52%)	1/33(3%)	<0.001**

ESR: Erythrocyte sedimentation rate; CRP: C-reactive protein; *Paired T Test;**Chi-Square test.

Conclusion: Methotrexate has so far been added to IGM treatment in order to decrease steroid dosage or to treat relapses, and it has been reported to be effective in case study and a limited number of studies with few patients [2, 4, 5]. Steroid + methotrexate treatment used by us in patients with IGM, which is a rare disease and for which no consensus exists regarding the treatment protocol, is effective and reliable in providing clinical improvement and long-term remission. Therefore, this treatment appears to be successful owing to long-term remission outcomes and very low relapse rates, without the patients having to undergo a surgical procedure and experience the associated anxiety and complications.

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AB1054 CLINICAL CHARACTERISTICS OF PATIENTS WITH UVEITIS FROM AN AUTOIMMUNE OCULAR DISEASE UNIT

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Background: The eye, particularly the uvea due to its substantial vascularization, can be a target for various immune reactions. Etiology is unknown in most cases of inflammatory ocular disease. Occasionally, ocular symptoms may be the first manifestation of some systemic diseases and its appearance, therefore, can lead us to the diagnosis and institution of treatment that, if performed early, can avoid irreversible sequelae.

Aim: Describe the experience at 12 months of an autoimmune ocular disease unit.

Objectives: to describe the clinical characteristics of patients diagnosed with uveitis and its association with systemic disease.

Methods: Retrospective, descriptive study. We included patients diagnosed with uveitis attending the Autoimmune Eye Disease Unit from January 2019 to December 2019. Qualitative variables were expressed as frequencies and percentages and quantitative variables as means and standard deviation.

Results: A total of 40 patients were included, 72.5% were women (26/40). Mean age of disease onset was 38 ± 17.2 years and the average diagnostic delay was 19.4 ± 46.8 months. The most frequent chief complaint was decrease in visual acuity, cited in 43.8% of the consults (25/57). Involvement was bilateral in 55% of cases (22/40). In order of decreasing frequency, diagnosis found were: Anterior uveitis in 47.5% (19/40), panuveitis in 35% (14/40), posterior uveitis in 10% (4/40) and intermediate uveitis in 7.5% (3/40). There was a recurrence (2 or more episodes of uveitis) in 65.7% (25/38) of patients and 58.8% (20/34) of them presented some sequelae. Prior to their first episode of uveitis, 12.5% (5/40) of patients had already been diagnosed with an autoimmune disease, the most common of which was spondyloarthritis (3/5). Meanwhile, 33.3% (13/39) were diagnosed with a systemic disease from their first episode of uveitis. Regarding laboratory tests, 46.4% (13/28) of patients presented acute phase

reactants, 13% (3/23) had positive serum antibodies and 41.6% (5/12) were HLA-B27 positive. Concerning treatment, 76.3% of patients required systemic corticosteroids (29/38) and 75% received at least one immunosuppressive drug (30/40). Out of this group, 30% needed a second immunosuppressive drug. Response to treatment was good in 63.6% of patients (21/33), partial in 18.1% (6/33), poor despite treatment in 6% (2/33) and poor due to lack of adherence in 12.1% (4/33).

Conclusion: In our cohort, there was a predominance of female, middle-aged patients with bilateral involvement. Anterior uveitis was the most frequent diagnosis. In one-third of patients, the first episode of uveitis led to diagnosis of a systemic disease. Most of our patients presented some type of sequel or local complication and required systemic treatment with corticosteroids and immunosuppressants.

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AB1055 CERTOLIZUMAB PEGOL: A SAFE AND EFFICIENT TREATMENT IN PATIENTS WITH UVEITIS DURING PREGNANCY.

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Background: Anti-TNFα agents are useful in uveitis (1-5). Certolizumab pegol (CZP) differs from other anti-TNFα agents due to its limited placental transfer.

Objectives: To assess efficacy and safety of CZP in women with uveitis during pregnancy.

Methods: Multicenter study of women with uveitis under CZP during pregnancy and their neonates.

Results: 14 women (23 eyes); mean age 34.3±5.5 yrs (TABLE 1). Pattern of uveitis: 10 anterior, 2 posterior, 1 intermediate, 1 panuveitis. Uveitis was bilateral in 9 and chronic in 7. CZP was started before conception in 10 patients and after in 4. All patients obtained or maintained ocular remission throughout pregnancy (FIGURE). Prednisone was reduced from a mean dose of 21.7±19.7 mg/day to 4.1±3.8 mg/day at 6 months (p=0.03), leading to complete discontinuation in 4. 15 healthy infants were born. Only 1 woman presented a mild infection. No infections or malformations were found in neonates after a follow-up of 6 months. 6 infants were breastfed and all received scheduled vaccinations without complications (TABLE 2).

TABLE 1.

	Age	Underlying disease	Immunosuppressants before CZP	Combined treatment
1	34	SpA	MTX, AZA, ADA	AZA
2	37	SpA	MTX, AZA, IFX, ADA, GOL	
3	39	SpA	AZA, ADA	AZA
4	46	SpA	CyA, ETN, ADA, IFX, GOL	
5	32	SpA	SSZ, ADA	SSZ
6	36	SpA	MTX, HCQ, ADA	
7	40	SpA	MTX, LFN, HCQ, IFX, ADA, GOL	HCQ
8	31	Idiopathic	MTX, MMF, CyA, ADA	
9	33	Idiopathic	MTX, AZA, ADA, ETN	
10	32	RA	MTX	AZA
11	23	Vogt-Koyanagi-Harada	AZA, ADA	AZA
12	36	Juvenil Idiopathic Arthritis	ADA	
13	32	Punctate inner choroidopathy	ADA	
14	29	Behcet	CyA, IFX, ADA	

TABLE 2.

	Full term pregnancy	Multiple gestation	Preconception CZP exposure	Labor complications	Maternal infections	Neonatal infections (< 6 m after birth)	Congenital malformations	Breast-feeding
Neonates, n/N	15/15	2/15	5/15	0/15	1/15	0/15	0/15	6/15

Conclusion: CZP seems to be effective and safe in female patients with uveitis during pregnancy and neonates.

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AB1056

SYMPTOMATIC SCLEROSING MESENTERITIS REVEALING ERDHEIM-CHESTER DISEASE: A RARE CONDITION MEDIATED BY BRAF

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Background: Sclerosing Mesenteritis (SM) refers to an entire spectrum of digestive inflammatory disorders. Diagnosis is based on imaging showing an increase of fat attenuation displacing bowel loops and is in most cases non-symptomatic. Several conditions (abdominal trauma/surgery, neoplasia, infectious and inflammatory diseases) are responsible for SM (1). Among neoplasia, Erdheim-Chester disease (ECD) is a rare clonal histiocytosis characterized by long bone involvement, peri-nephric fat infiltration and cardio-vascular involvement associated with compatible histology (2). Biopsy is mandatory to confirm tissue infiltration by histiocytes and detect somatic mutation. Almost 80% of ECD patients harbor mutation in mitogen activated protein (MAP) kinase pathway especially *BRAF*^{V600E}

gene mutation in about 60% of cases (3). No series of patients presenting both pathologies has been reported. Furthermore, no correlation with *BRAF* mutation status has been described in patient harboring SM and ECD.

Objectives: To describe the clinical, radiological and mutational status of patients harboring SM and ECD.

Methods: We reviewed the database of patients with histiocytic disorders in Besancon University Hospital. Patient required one abdominal computed tomography showing sclerosing mesenteritis and clinical/histological features of ECD to fulfill the inclusion criteria. All biopsy samples were investigated for mutation of MAP kinase pathway gene.

Results: Four patients suffered from SM and ECD. The median age at the diagnosis of ECD was 68 years old (61-72). All patients described abdominal pain and the mean duration between first symptoms and diagnosis of ECD was 12 months (4-19). The mean CRP level at diagnosis was 40.75 mg/L (5-117). Two patients were found to have myeloid neoplasms (chronic myelomonocytic leukemia (#2) and essential thrombocythemia (#4)) concurrent with ECD diagnosis.

Regarding abdominal computed tomography, all patients had a mesenteric mass associated with hyper-attenuated mesenteric fat and a "fat halo sign". One patient (#2) had ascites and one had splenomegaly (#4) but no patient had enlarged lymph nodes. CT also demonstrated peri-nephric fat infiltration ("hairy kidney") (4/4), vascular sheathing of aortic branches (3/4), adrenal hypertrophy (1/4) or ureter dilation (1/4). The mean SUV_{max} of the mesentery was 7.5 (4.1-10.9) at diagnosis on ¹⁸F- fluorodeoxyglucose-PET. Three patients underwent mesenteric fat biopsy and all samples exhibited ECD histology. Regarding mutational status, 75% (3/4) patients had *BRAF*^{V600E} mutation.

After initiation of therapies for ECD (targeted therapies for 3/4 patients), all patients had improvement of digestive symptoms and decreased of SUV_{max} on evaluation ¹⁸ FDG-PET during the follow up.

Conclusion: ECD should be investigated in patient with symptomatic SM especially if it is associated with peri-nephric fat infiltration. This condition is rare and might be driven by BRAF gene.

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AB1057

SCHNITZLER'S SYNDROME IN THE DIFFERENTIAL DIAGNOSIS OF ADULT STILL'S DISEASE

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Background: Schnitzler's syndrome (SchS) and adult onset Still disease (AOSD) are currently considered as multifactorial autoinflammatory diseases (MAIDs) and are classified as systemic inflammation with urticarial rash. Clinical similarities between SchS and AOSD (fever, urticarial rash, arthralgias), increased ESR and CRP and the efficacy of IL-1 inhibitors may lead to the diagnostic delay in SchS pts. Testing for monoclonal gammopathy helps establish the diagnosis in SchS pts but is not routinely used in AOSD pts.

Objectives: to examine demographic, clinical and laboratory characteristics, and the therapy of SchS pts in a single rheumatology center.

Methods: 5 SchS patients (2 females, 3 males), aged 32 to 68, underwent inpatient and outpatient examinations in the rheumatology center. All pts underwent a standard rheumatology examination, including ESR, CRP and M-gradient. 4 pts underwent genetic testing for mutations in *NLRP3*, *TNFRSF1A* genes to exclude MAIDs, such as CAPS and TRAPS.

Results: All pts were initially diagnosed with AOSD. The age at onset ranged between 28 and 66 years. Time to diagnosis varied from 2 to 22 years, being within 4 years in 4 of 5 pts. Patients presented with fever (4), urticarial rash (5) and musculoskeletal manifestations (5) (arthralgia in 3, bone pain in 4). Of 2 pts with serositis one presented with pericarditis and another – with pleuritis. Only 1 demonstrated a sore throat and polyneuropathy of the lower extremities. ESR and CRP were increased in all pts, leukocytosis was noted in 4 (Table 1). The