SAT0625

SARCOIDOSIS IS ASSOCIATED WITH AN INCREASED RISK OF GASTROINTESTINAL EVENTS: A POPULATION-BASED RETROSPECTIVE COHORT STUDY 1976–2013

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Background: An increased risk of gastrointestinal (GI) diseases has been observed in several chronic inflammatory disorders but the risk in patients with sarcoidosis is not known.

Objectives: This study was undertaken to characterise the risk of GI diseases in patients with sarcoidosis.

Methods: A population-based cohort of 345 incident cases of sarcoidosis among Olmsted County, Minnesota residents in 1976–2013 was identified from a comprehensive medical record-linkage system. Diagnosis was confirmed by individual medical record review. A cohort of 345 sex and age-matched comparators was also identified from the same underlying population. Medical records of both groups were reviewed for GI diseases. Cox models adjusted for age, sex and calendar year were used to compare the rate of development of GI diseases between the groups.

Results: GI events occurred in 101 cases and 63 comparators, corresponding to an adjusted hazard ratio (HR) of 1.90 (95% confidence interval [CI] 1.38–2.61). Patients with sarcoidosis had an increased risk for both upper (HR 1.90; 95% CI 1.27–2.83) and lower GI events (HR 1.97; 95% CI 1.27–3.05) relative to comparators. By disease type, patients with sarcoidosis had a significantly elevated risk of upper GI ulcer, upper GI haemorrhage and diverticulitis (**table 1**).

Comorbidity	Number of events prior to index date	p-value	Number of events after index date	Cumulative incidence at 10 years for non-sarcoidosis subjects (±SE)	Cumulative incidence at 10 years for sarcoidosis patients (±SE)	Hazard ratio (95% CI)	p-value
Upper GI diagnoses							
Upper ulcer	21/19	0.87	11/27	1.9 (0.2, 3.6)	4.5 (2.0, 6.8)	2.56 (1.27, 5.17)	0.009
Upper bleed	6/14	0.11	14/29	0.9 (0.0, 2.0)	4.4 (1.9, 6.9)	2.36 (1.24, 4.47)	0.009
Upper perforation	2/0	0.50	0/1	–	–	–	–
Upper obstruction	1/3	0.62	7/8	1.4 (0.0, 2.7)	0.3 (0.0, 0.9)	1.20 (0.43, 3.32)	0.72
Esophagitis	7/4	0.55	18/27	3.6 (1.4, 5.8)	5.4 (2.7, 8.1)	1.58 (0.87, 2.88)	0.13
Esophageal varices	0/3	0.25	1/6	0.0 (0.0, 0.0)	0.7 (0.0, 1.8)	6.51 (0.78, 54.16)	0.083
Lower GI diagnoses							
Lower ulcer	2/1	1.0	2/7	0.6 (0.0, 1.4)	1.4 (0.0, 2.8)	3.51 (0.73, 16.92)	0.12
Lower bleed	5/3	0.72	12/15	1.8 (0.2, 3.3)	2.9 (1.0, 4.8)	1.32 (0.62, 2.82)	0.48
Lower perforation	0/0	–	2/1	–	–	–	–
Lower obstruction	0/2	0.50	1/2	–	–	–	–
Diverticulitis	3/2	1.0	9/18	0.4 (0.0, 1.1)	2.3 (0.6, 4.0)	2.29 (1.03, 5.10)	0.043
Ischaemic colitis	2/0	0.50	3/7	0.0 (0.0, 0.0)	0.6 (0.0, 1.5)	2.38 (0.61, 9.26)	0.21
Infectious colitis	1/5	0.22	7/13	0.6 (0.0, 1.5)	2.9 (0.9, 4.9)	1.99 (0.79, 5.00)	0.14

Conclusions: Patients with sarcoidosis have a higher risk of both upper and lower GI events compared with subjects without sarcoidosis.

Disclosure of Interest: None declared

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ANAKINRA TREATMENT IN REFRACTORY CASES OF ADULT-ONSET STILL DISEASE: CASE SERIES

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Background: Despite methotrexate and steroid treatment, in cases of Adult-onset Still's disease (AOSD) it is usually difficult to maintain clinic stability. In refractory cases, Anakinra treatment has been reported to be efficacious.¹

Objectives: In this retrospective review, it is aimed to evaluate the AOSD cases treated with anakinra in our centre.

Methods: Fourteen AOSD patients (11 female, 3 male) who were being followed in our outpatient clinic were reviewed retrospectively. The demographic characteristics, pre- and post-treatment clinical findings were reported.

Results: The mean follow-up period of the patient population was 33.5±30.07 months (mean ±SD). Initial prednisolone dose was 37.3 mg/day. Except for one, all of our patients were exposed to methotrexate before being treated with anakinra. This patient was being treated with cyclosporine instead, since she had concomitant Macrophage Activation Syndrome. The other medications, the patients were previously treated with, were Etanercept (n=2), Tocilizumab (n=3), Infliximab (n=1) and Adalimumab (n=1).

All patients were on 100 mgs of Anakinra, daily, except for the one treated with 200 mg/day. The mean duration of Anakinra therapy was 11.4 months. Among 7 patients in whom anakinra therapy was terminated, 1 had drug induced urticaria, 1 was primary irresponsive, 4 were secondary irresponsive and the other had severe pneumonia. Primary irresponsiveness is the lack of response to the therapy since the drug was first introduced, whereas in secondary irresponsiveness the case responds to the medication for a while and starts to flare again after asymptomatic-free period on the medication. Among 14, 7 of our patients are still on 100 mg/d Anakinra.

The mean level of C reactive protein (CRP) measures was reduced from 64.38 ±61.95 mg/L to 34.3±24.3 mg/L with Anakinra therapy (p=0.003). Similarly, mean Erythrocyte Sedimentation Rate (ESR) was dropped to 33±22 mm/h from 59 ±35 mm/h by the help of the therapy (p<0.001). Among patients who primarily responded Anakinra therapy the mean Ferritin measures dropped to 427.25 ng/ml from 910 ng/ml (p=0.006). On the other hand, the Ferritin level was not significantly reduced in patients who did not respond Anakinra. The mean Patient reported Global Visual Analogue Scale (PG-VAS) score was also decreased to 3.83±4.7 from 9.5±0.07 following the therapy (p<0.001). Unfortunately, one of our 7 patients who were followed in remission under Anakinra died of an unknown etiology.

Conclusions: Adult-onset Still's disease is a challenging disorder, lacking a sufficient long-time clinical control. In order to obtain a full remission, various efforts have been spent so far. One of these approaches is to treat refractory cases with Anakinra, an IL-1 blocking agent. According to our clinical experience we state that, anakinra has a relatively high efficacy in controlling refractory cases.

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SAT0627

FABRY DISEASE: DIAGNOSTIC ERRORS IN RHEUMATOLOGY PRACTICE

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Background: Fabry disease (FD) is a rare X-linked storage disease resulting from the deficient activity of the lysosomal α-galactosidase A (AGAL) and leading to a progressive accumulation of glycosphingolipids in a wide range of cell types throughout the body, including kidney, heart and nervous system. Certain manifestations of FD can mimic those of rheumatic diseases.

Objectives: To evaluate the occurrence and the possible causes of diagnostic errors in patients with FD in rheumatology practice.

Methods: We retrospectively studied the medical records of 82 adult patients with definite FD (low or absent AGAL activity, pathogenic mutation in the GLA gene, typical clinical symptoms in patient and/or his relatives, results of kidney biopsy). There were 55 males and 27 females aged of 18 to 69 years (median 38²⁹, 48⁴⁸ years).

Results: Seventy two of 82 patients (87.8%) had a history of classic phenotype of FD from childhood or adolescence. The typical manifestations included neuropathic pain that was related to heat and fever (65/82; 79%), angiokeratoma (37/