Table 1. MAS-SALIENT FEATURES

FEATURES	INITIAL MANIFESTATIONS [n=8]	KNOWN RHEUMATIC DISEASES[n=8]	P VALUE (<0.05)
Primary rheumatic diseases			
SLE	5(62.5%)	7(87.5%)	-
AOSD	2(25%)	1(12.5%)	
Kikuchi disease	1(12.5%)	0	
Fever	8(100%)	7(87.5%)	0.30
Respiratory	2(25%)	5(62.5%)	0.13
Neurological	1(12.5%)	4(50%)	0.10
Bleeding	2(25%)	1(12.5%)	0.52
Splenomegaly	8(100%)	7(87.5%)	0.30
Hepatomegaly	6(75%)	3(37.5%)	0.13
Haemoglobin(g/dl)	8.58(5.4-11.7)	8.2(5.2-11.5)	0.70
WBC count(μ L)	5550(1000-11800)	34775(1200-6200)	0.19
Platelet count($\times 10^9$ /l)	1.41(0.30-2.4)	1.35(0.32-1.8)	0.90
ESR(mm/hr)	76.75(42-137)	68(20-127)	0.65
AST(U/L)	666(36-3481)	257.75(45-642)	0.36
ALT(U/L)	252.5(18-790)	217.62(28-1044)	0.82
LDH(U/L)	2490.12(164-16,000)	938.75(357-1569)	0.44
Ferritin(ng/ml)	2187.46(532-7500)	3754.75(531-12536)	0.37
Triglycerides(mg/dl)	316(53-551)	275.8(160-317)	0.52
Fibrinogen(mg/dl)	129.8(76-178)	200.62(140-292)	0.009
Complement 3(mg/dl)	39.14(13-60)[n=5]	57.72(32-95)[n=7]	0.16
Complement 4(mg/dl)	4.24(1.2-6)[n=5]	14.22(2.8-34)[n=7]	0.07
Anti dsDNA antibodies[ELISA] IU/ml	596.2(90-1151)[n=5]	316.82(10-800)[n=7]	0.26

MAS-Macrophage activation syndrome, WBC-white blood cell, ESR-erythrocyte sedimentation rate, AST-aspartate aminotransferase, ALT-alanine aminotransferase, LDH-lactate dehydrogenase, dsDNA-double stranded deoxyribonucleic acid

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.2965

AB0774

TREATMENT WITH IL-1 INHIBITORS IN PATIENTS WITH SCHNITZLER'S SYNDROME: A 5-YEAR CLINICAL EXPERIENCE

S. Salugina¹, V. Gorodetskiy², E. Fedorov¹, V. Rameev³, E. Borzova^{4,5}, N. Lopatina⁶, ¹V.A.Nasonova Research Institute of Rheumatology, Pediatric Department, Moscow, Russian Federation; ²V.A.Nasonova Research Institute of Rheumatology, Laboratory of Intensive Therapy Methods, Moscow, Russian Federation; ³Sechenov's First Moscow State Medical University, E. M. Tareev Clinic of Rheumatology, Neurology and Occupational Pathology, Moscow, Russian Federation; ⁴Sechenov's First Moscow State Medical University, Department of Dermatology and Venereology, Moscow, Russian Federation; ⁵Research and Clinical Institute of Pediatrics n.a. Yuri Veltischev, Department of Clinical Genetics, Moscow, Russian Federation; ⁶V.A.Nasonova Research Institute of Rheumatology, 5th Rheumatology Department with Hematology Beds, Moscow, Russian Federation

Background: Schnitzler's syndrome (SchS) is a rare non-monogenic auto-inflammatory disease (AID) which manifests in adults and is characterized by recurrent urticarial lesions, monoclonal gammopathy, and other manifestations of a systemic inflammatory response (fever, muscle / joint / bone pains, leukocytosis, increased ESR, CRP). Off-label treatment with IL-1 inhibitors (anakinra, canakinumab) was shown to be highly effective and well tolerated in pts with SchS based on a few multicenter studies and practical experience.

Objectives: To analyze a real-life experience of the treatment with IL-1 inhibitors in pts with SchS in the rheumatology tertiary care settings.

Methods: The study included 7 pts (3 women, 4 men) with treated with IL-1 inhibitors (anakinra, canakinumab). A median age at the diagnosis was 44 years old (25 – 68). Age at the disease onset ranged from 22 to 66. All pts met the Strasbourg diagnostic criteria. 7 pts received canakinumab, while 3 pts were first treated with anakinra followed by a switch to canakinumab. Anakinra was administered subcutaneously 100mg per day. Canakinumab was given subcutaneously at a dosage of 150mg every 8 weeks. The treatment duration with anakinra ranged from 1 week to 2.5 months with further switching to canakinumab. The duration of canakinumab treatment by the time of analysis ranged from 2 months to 5.5 years.

Results: All pts at the inclusion into the study suffered from urticarial rash, pains in bones and joints, fatigue, fever (6), increased acute phase marker levels (ESR, CRP), monoclonal gammopathy, mainly IgMk (4.7 – 9g/l). An average period between the disease onset and the treatment initiation was 3 years (2 to 4). In all pts lymphoproliferative diseases and monogenic AIDs (CAPS, TRAPS) were excluded. Before the treatment with IL-1 inhibitors, all pts had received NSAIDs and/or GCs, methotrexate - 2, plaquenil-2. During the treatment with IL-1 inhibitors, 6 out of 7 pts had a complete response as demonstrated by a symptom resolution within several days and a rapid decrease in ESR and CRP levels. In 3 pts, who responded anakinra, the treatment was switched to canakinumab. In one

patient with a partial response to anakinra, there was a loss of the effect upon switching to canakinumab. In one patient, the interval between canakinumab injections was increased up to 5 months without a symptom relapse. This patient fathered a healthy child whilst on treatment with canakinumab. The therapy was well tolerated in all pts, no SAE were noted.

Table 1. Clinical and demographic characteristics of patients with SchS

Patients	1	2	3	4	5	6	7
Sex	M	F	M	F	F	M	M
Age (y)	44	32	56	68	25	51	49
Age of disease onset (y)	40	29	53	66	22	39	47
Urticarial rash	+	+	+	+	+	+	+
Fever	+	-	+	+	+	+	+
Fatigue	+	+	+	+	+	+	+
Bone pain	+	+	+	+	+	+	+
Arthralgia/arthritis	+	+	+	+	+	+	+
ESR(normal <15mm/h)	31	140	100	49	81	53	56
CRP (normal <5mg/l)	107	44	192	96	369	76	41,82
IgM kappa (g/l)	7,1-5,2	4,7-7,8	7,6	-	-	5,2	9
IgM lambda (g/l)	-	-	1,36	-	-	-	-
IgG kappa (g/l)	-	-	-	5,1	-	-	-
IgG lambda (g/l)	-	-	-	-	10,7	-	-

Conclusion: SchS is a rare disease that occurs in the practice of a rheumatologist. Short and long acting IL-1 inhibitors are highly efficient and safe treatment options for pts with SchS. A switching from short to long acting IL-1 inhibitors can be effective in SchS pts.

REFERENCES:

- [1] Krause K, Tsianakas A, Wagner N, Fischer J, Weller K, Metz M, et al. Efficacy and safety of canakinumab in Schnitzler syndrome: a multicenter randomized placebo-controlled study. *J Allergy Clin Immunol* 2017;139:1311-20.
- [2] Betrains A., Staels F, Vanderschueren S. Efficacy and safety of canakinumab treatment in schnitzler syndrome: A systematic literature review *Seminars in Arthritis and Rheumatism* 50 (2020) 636-642.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3176

AB0775

MANY RARE AND SERIOUS LOCALIZATIONS IN ONE CASE WITH HEREDITARY EXOSTOSES

R. Sarraj¹, M. Ghali¹, R. Ben Chehida¹, G. Mouna¹, I. Bejia¹, J. Mahbouba¹, S. Zrour¹, M. Touzi¹, N. Bergaoui¹. ¹Fattouma Bourguiba Hospital, Rheumatology, Monastir, Tunisia

Background: Multiple hereditary exostoses (MHE) is an inherited disorder of bone growth. People who have MHE grow exostoses or bony bumps on their bones that vary in size, location and number. Some localizations are rare but potentially serious such as ribs, spine or pelvis due to proximity to important structures.

Objectives: The aim of this case report was to illustrate the clinical and radiographic features of some rare localizations of MHE.

Methods: We are reporting a rare case of a thirty one-year-old male with severe localizations of MHE.

Results: A thirty one-year-old male affected by hereditary multiple exostoses., not followed up, came to our consultation complaining about chest pain. On examination, he walks with a limp because of the unequalness of his limbs. The osteo-articular examination revealed numerous palpable osseous masses at the distal and proximal end of the 2 arms, 2 forearms, 2 thighs and 2 legs and at the right iliac wing. Neurological examination revealed vivid deep tendon reflexes, bilateral positive Hoffmann's sign and Babinski reflex associated with right hemiparesis. The pulmonary exam was normal. Medullary MRI showed medullary compression with spinal cord atrophy and sign of myelopathy of c1 c2 segment. The ct – scan revealed multiple exostosis, the two largest lesions were located in the second rib measuring 45mm * 35mm axially and 52mm in height compromising blood vessels, and in the right iliac bone with a retroperitoneal growing mass measuring 126 * 94 mm axially and 100mm in height leading to a serious entrapment of the axial skeleton and the abdominal organs. It also confirmed thickening of the cartilage cap of the right iliac bone suggestive of malignant chondrosarcomatous transformation.

Conclusion: Exostoses are the most common benign bone tumors. Its complications are rare but can cause a functional disability or even a vital risk.

REFERENCES:

- [1] Eke GK, Omonakwe HE, Echem RC. Hereditary multiple exostoses in a 15-year-old boy: A case report and review of literature. *Niger J Paediatr*. 2016;43(4):295.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3296