Abstract 128 Table 1 Patients characteristics and associations with LSI by disease duration, at baseline

Variables *	Descriptive Statistics **	Association with LSI ***	
Patient's characteristics		Incident cases	Prevalent cases
Age	49.0 (36.8–58.5)	–0.19 (p=0.028)	–0.19 (p<0.001)
Gender		p=0.007	p=0.006
Women	589 (92)	5.4 (4.9–6.3)	6.3 (5.7–8.0)
Men	55 (9)	6.7 (5.6–8.3)	7.1 (5.8–8.5)
Ethnicity (n=592)		p=0.032	p=0.005
Caucasian	437 (74)	5.4 (4.7–6.4)	5.7 (5.1–7.9)
Asian	80 (14)	6.1 (5.4–8.5)	7.7 (5.3–8.2)
Other	75 (13)	5.9 (5.3–8.1)	5.9 (5.2–8.2)
Marital Status (n=586)		p=0.318	p=0.076
Single	107 (18)	5.7 (5.3–7.3)	6.5 (5.3–8.2)
Married	387 (66)	5.4 (4.7–6.3)	5.8 (5.2–8.0)
Other	92 (16)	5.4 (4.8–7.1)	5.7 (4.9–8.2)
Site		p=0.789	p=0.383
Winnipeg	231 (36)	5.8 (4.9–7.0)	5.6 (5.0–7.8)
Hamilton	25 (4)	6.2 (5.4–6.6)	6.6 (5.4–7.1)
Quebec	129 (20)	5.4 (4.6–6.8)	6.2 (5.2–8.3)
Ottawa	4 (1)	6.1 (6.1–6.1)	6.3 (5.7–8.2)
London	6 (1)	7.2 (7.2–7.2)	6.1 (5.5–7.8)
Calgary	244 (38)	5.4 (5.0–6.3)	5.9 (5.1–8.1)
Disease's characteristics			
Disease duration (n=623)	10.1 (2.7–20.6)	0.11 (p=0.241)	0.07 (p=0.106)
Recent diagnosis (<=15 months)		NC	NC
Ancient diagnosis (>15 months)			
ACR Criteria Score (n=626)	5 (4–6)	0.29 (p<0.001)	0.30 (p<0.001)
ACR LSI Score (n=626)	5.8 (5.1–7.9)	NC	NC
SLICC Criteria Score (n=538)	6 (5–8)	0.49 (p<0.001)	0.42 (p<0.001)
SLEDAI score (n=598)	2 (0–6)	0.05 (p=0.586)	–0.00 (p=0.992)
SLAQ Score (n=407)	12 (7–18)	–0.26 (p=0.023)	–0.18 (p=0.001)
SDI Score (n=609)	0 (0–2)	p=0.296	p=0.005
0	310 (51)	5.5 (4.8–6.4)	5.6 (4.9–7.9)
>0	299 (49)	5.5 (5.2–8.0)	6.2 (5.2–8.2)
Medication			
Prednisone (n=614)		p<0.001	p<0.001
Yes	203 (33)	6.4 (5.3–8.2)	6.8 (5.5–8.2)
No	411 (67)	5.4 (4.7–6.0)	5.6 (4.9–7.7)
Prednisone dose (n=219)	7.5 (5.0–15.0)	0.41 (p=0.001)	–0.01 (p=0.92)
Immunomodulators (n=616)		p=0.189	p=0.028
Yes	554 (90)	5.5 (5.0–6.9)	5.9 (5.2–8.1)
No	62 (10)	5.3 (4.4–6.2)	5.5 (4.9–6.4)
Antimalarial		p=0.241	p=0.97
Yes	517 (83)	5.5 (4.9–6.8)	5.8 (5.1–8.1)
No	106 (17)	5.2 (4.6–6.3)	5.8 (4.9–8.2)
Biologic Agent (n=609)		p=0.800	p=0.270
Yes	25 (4)	5.9 (5.3–6.5)	6.1 (5.3–8.4)
No	584 (96)	5.5 (4.9–6.8)	5.8 (5.1–8.1)
Death during follow-up		NC	p=0.002
Yes	12 (2)		8.2 (7.0–9.3)
No	627 (98)		5.8 (5.1–8.1)

*n=639 except when specified; **Results presented as Median (1 st quartile 3rd quartile) or Frequency (percentage) for continuous and categorical variables respectively; ***Results presented as Spearman Correlation coefficient (pvalue) or Median (1 st quartile 3rd quartile) with Kruskal-Wallis pvalue for continuous and categorical variables respectively; NC: Not calculated.

like disease activity and prednisone dose - modifiable. LSI can predict adverse outcome such as damage or death over time.

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MACROPHAGE ACTIVATION SYNDROME AS A PRESENTATION IN PEDIATRIC LUPUS: A RETROSPECTIVE STUDY OF 3 CASES

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Background Macrophage activation syndrome (MAS) can, at times, be the presentation of pediatric lupus and diagnosis requires high index of suspicion.

Methods We retrospectively studied 140 pediatric lupus patients from January 1993- November 2018 and collected clinical and laboratory data of patients (3) who had MAS as presenting manifestation.

Results Case 1 was 11-year-old girl with fever for 4 months associated with rash and generalized body swelling for 1 month. Examination revealed rash over malar area and ear lobules, anasarca, hepatomegaly, bilateral pleural and pericardial effusion. In view of multisystem involvement a possibility of lupus was considered which was confirmed by investigations (table 1). She had elevated ferritin and fasting triglyceride and low fibrinogen. A clinical possibility of lupus with associated MAS was considered. She received pulses of methylprednisolone, one dose of intravenous immunoglobulin following which she improved. In view of nephrotic range proteinuria she was started on induction regimen with cyclophosphamide and shifted to mycophenolate in maintenance. Her initial SLEDAI-2k was 32- this decreased to 4 at 3 year follow-up.

Case 2 was a 9-year-old girl with fever, rash, generalized body swelling for 1 month and altered sensorium for 4 days. Examination revealed pallor, oral ulcers and hepatomegaly. She was in shock at presentation. In view of multisystem involvement a possibility of lupus was considered which was confirmed by investigations (table 1). She had pericardial effusion and low ejection fraction (25%). A possibility of MAS was considered and investigations revealed hyperferritinemia, elevated triglyceride and hypofibrinogenemia. She was given methylprednisolone pulses and continued on oral prednisolone, mycophenolate and hydroxychloroquine. Her initial SLEDAI-2k was 17- this decreased to 0 at 3 year follow-up.

Case 3 was an 8-year-old girl who had fever, rash and body swelling for 15 days. She had tachycardia, tachypnea, pallor, anasarca, subconjunctival bleed and frontal alopecia. She had pleural and pericardial effusion. In view of multisystem involvement a possibility of lupus was considered which was confirmed by investigations (table1). She had high ferritin and triglyceride. So a possibility of MAS was considered and she received pulse methyl prednisolone and intravenous immunoglobulin. She also had hematuria and proteinuria (renal biopsy could not be performed as she was sick and had thrombocytopenia) so was given pulse cyclophosphamide

Abstract 129 Table 1

Investigations	Case 1	Case 2	Case 3
Haemoglobin (g/L)	70	67	70
White cell counts (/L)	1.5×10 ⁶	2.5×10 ⁶	3.5×10 ⁶
Lymphocyte count (/L)	0.4×10 ⁶	0.6×10 ⁶	0.8×10 ⁶
Platelet count (/L)	63×10 ⁹	50×10 ⁹	76×10 ⁹
Urine protein (mg/m ² /hour)	74		
C3 (Normal 50–150 mg/dL)	<27	<27	<27
C4 (Normal 20–50 mg/dL)	4	3	3
ANA (immunofluorescence method)	4+diffuse	3+diffuse	4+diffuse
Anti dsDNA (N:<60 IU/mL)	707	583	1394
Serum ferritin (ng/ml)	3233	6767	1019
Fasting triglyceride (mg/dl)	442	504	328
Fibrinogen (mg/dl)	120	160	

followed by mycophenolate. Her initial SLEDAI-2k was 25- this decreased to 0 at 2 year follow-up.

Conclusions MAS can be the presenting manifestation of pediatric lupus and may contribute to disease severity and requires aggressive management

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JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS RELATED PANCREATITIS: AN UNCOMMON MANIFESTATION OF A COMMON DISEASE

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Background Pancreatitis is a rare but potentially life-threatening complication of systemic lupus erythematosus(SLE).SLE-related pancreatitis can be a presenting manifestation of SLE or may occur during follow-up.

Methods We have reviewed the clinical records of 140 children with SLE between period of 1993–2018.We report 3 children with SLE who presented with acute pancreatitis.

Results Case1–12-year-girl presented with fever and alopecia. Examination revealed pedal oedema,periorbital puffiness,large joint arthritis.Investigations showed anemia,thrombocytopenia, elevated erythrocyte sedimentation rate(ESR),normal renal functions,microscopic hematuria,nephrotic range proteinuria, hypocomplementemia,antinuclear antibody (ANA) 4+,elevated anti double stranded DNA (anti-ds DNA),negative antiphospholipid antibodies (APLA) titre(table).Renal biopsy revealed Class 3 lupus nephritis and initiated on intravenous methylprednisolone.Two days later,she developed severe epigastric pain and vomiting.The pain increased in severity and physical examination revealed marked tenderness in the epigastrium. Serum amylase and lipase were elevated.Clinical possibilities included steroid induced pancreatitis and lupus pancreatitis.In view of severity of symptoms,intravenous methylprednisolone was continued following which she showed a dramatic improvement and normalisation of pancreatic enzymes.There has been no recurrence of pancreatitis over 3 years of follow-up period and remained well on low dose oral prednisolone (5 mg),mycophenolate-mofetil and hydroxychloroquine.

Abstract 130 Table 1 Investigation Table

Investigation	Case 1	Case 2	Case 3
Haemoglobin (g/L)	92 g/L	96 g/L	80 g/L
White cell counts	7.7×10 ⁶ /L	7.8×10 ⁶ /L	7.5×10 ⁶ /L
Lymphocyte count	2.15×10 ⁶ /L	1.9×10 ⁶ /L	1.95×10 ⁶ /L
Platelets	L	L	L
Urine routine	130×10 ⁹ /L	150×10 ⁹ /L	420×10 ⁹ /L
Urine protein (mg/m ² /hour)	L	L	L
Urine routine	10–12	Few RBC,	No RBC,
Urine protein (mg/m ² /hour)	RBC, 3	3+albumin	3
	+albumin	82 mg/m ² /hr	+albumin
			40 mg/m ² /hr
C3 (Normal 50–150 mg/dL)	23.4 mg/dl	129 mg/dL	34 mg/dl
C4 (Normal 20–50 mg/dL)	2.98 mg/dl	37 mg/dL	10 mg/dl
ANA	4+diffuse	3 +	3+diffuse
Anti dsDNA (N:<60 IU/mL)	890	<60	123
Antiphospholipid antibodies: a)	Negative	Negative	Positive
Lupus anticoagulant b)	Negative	Negative	Negative
Anticardiolipin antibody (IgG and IgM) c) Anti B2 Glycoprotein –1 antibody (IgG and IgM)	Negative	Negative	Negative
Skin biopsy	Not done	Positivity of lupus band test with high positive IgG, IgA, IgM and C3 in dermal vessels	Lupus band test -positive
Renal biopsy	Class 3 lupus nephritis	IgG, Ig A, Ig M positivity in the mesangium as well as capillary loops and C3 in small sized blood vessels	Class 4 lupus nephritis
Serum amylase (<100 U/L)	238 U/L	400 U/L	290 U/L
Serum lipase (<60 U/L)	231 U/L	Not done	Not done

Case2-A-6-year-old presented with pain abdomen and vomiting.Physical examination showed epigastric tenderness.Investigations showed anemia,thrombocytopenia,elevated amylase levels(table).Computerised tomography(CT) abdomen revealed acute necrotising pancreatitis.A follow-up ultrasound abdomen revealed a pancreatic pseudocyst.He had a second episode of acute pancreatitis along with anasarca after 3 months which improved with conservative management.Investigation showed anemia,nephrotic range proteinuria and microscopic haematuria(table).ANA was positive with normal complement levels and negative anti-dsDNA titres(table).Lupus band test on skin biopsy was positive.Renal biopsy showed mesangioproliferative-glomerulonephritis with full house pattern.Following the initiation of steroids,he improved and there has been no recurrence of pancreatitis over the next 4 years.

Case 3–9 year-girl presented with generalised rash,alopecia for 5 months.She also had pain abdomen for last 2 months. Investigations showed elevated amylase,ultrasound abdomen revealed acute pancreatitis.She had undergone a laparotomy elsewhere.Examination showed generalised pigmented rash,alopecia, periorbital puffiness,hard palatal ulcer,surgical scar on the abdomen.Investigations showed anemia, normal platelet count,elevated ESR,normal renal functions,nephrotic range proteinuria,elevated serum amylase.Ultrasound and CT abdomen revealed a pancreatic pseudocyst.A clinical possibility of SLE