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

Funding Source(s): None

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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

with acute pancreatitis and lupus nephritis was considered. Further investigations were suggestive of lupus (table). Workup for APLA revealed positive lupus anticoagulant. Her skin biopsy was consistent with lupus and renal biopsy revealed class 4 lupus nephritis. She was initiated on oral prednisolone and was given pulses of intravenous cyclophosphamide. There has been no recurrence of pancreatitis over 12 years follow-up.

Conclusions Pancreatitis can at times, be the presentation of childhood lupus and requires prompt and aggressive management.

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STIMULATION OF MONONUCLEAR CELLS THROUGH TOLL-LIKE RECEPTOR 9 INDUCES RELEASE OF MICROVESICLES EXPRESSING DNA AND GALECTIN 3-BINDING PROTEIN IN AN INTERFERON-DEPENDENT MANNER

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Abstract 131 Figure 1 PBMCs from healthy donors (n=6) were incubated for 24 hours with the TLR9-agonist ODN2395, alone (-) or in combination with the IFN- inhibitor IFN alpha-IFNAR-IN-1 hydrochloride (+). MVs released into the culture supernatants were subsequently isolated by differential centrifugation, quantified and characterized with respect to expression of G3BP and DNA by flow cytometry. (A) Concentration in the culture supernatant of G3BP-expressing MVs, DNA-expressing MVs, and G3BP/DNA double-positive MVs. (B) Corresponding concentration of MVs expressing these markers when co-stimulating T-cells with anti-CD3 antibody. (C) The G3BP-derived median fluorescence intensity (MFI) of the G3BP-expressing MV population and (D) the DNA-derived MFI of the DNA-expressing MV population released from the ODN2395-treated PBMCs in presence (black columns) or absence (gray columns) of anti-CD3 antibody (OKT3). Columns and error bars represent median values and interquartile range. (*) P<0.1. *P<0.05.