European Journal of
Medical Research and Clinical Trials

Case Report Open Access

Acute Pancreatitis as initial Presentation of Paediatric Systemic Lupus Erythematosus

Ellen Ferdous*, Manik Kumar Talukder, Mohammad Imnul Islam, Mohammed Mahbubul Islam and Shahana A Rahman

Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

***Corresponding authors:** Ellen Ferdous, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh, Tel: 01711440498, E-mail: ellenferdous35@gmail.com

Received Date: June 10, 2021 **Accepted Date:** July 10, 2021 **Published Date:** July 12, 2021

Citation: Ellen Ferdous (2021) Acute Pancreatitis as initial Presentation of Paediatric Systemic Lupus Erythematosus. Eur J Med Res Clin Trials 1: 1-12.

Abstract

Systemic lupus erythematosus (SLE) is a chronic inflammatory autoimmune disease characterized by multisystem involvement. Although gastrointestinal symptoms are common, acute pancreatitis is a rare entity in Paediatric systemic lupus erythematosus (pSLE). Acute pancreatitis may occur in different pSLEdisease states, but its occurrence as an initial presentation is uncommon. Here, we report a 13-year-old girl who presented with severe epigastric pain and vomiting for two days. She also had an irregular fever, facial rash,polyarthralgia, oral ulceration, alopecia and cervical lymphadenopathy. Investigations showed elevated pancreatic enzymes with edematous pancreas.Other laboratory work-up diagnosed the girl as a case of pSLE.

Keywords: Acute pancreatitis; pSLE (Paediatric Systemic Lupus Erythematosus)

Introduction

Systemic Lupus Erythematosus is a multisystem autoimmune disease with protean clinical presentations. Almost every organ and system can be affected in SLE [1].Paediatric SLE (pSLE) hasa more aggressive disease course with widespread organ involvement than adult-onset SLE. Renal, neurological, hematological involvement and constitutional features are the usual presentations of pSLE [2].Gastrointestinal systemis also affected in pSLE patients. The incidence of gastrointestinal involvement was 19% in pSLE and 8-40% in adult SLE cases [3]. Acute Pancreatitis is anuncommon manifestation and its prevalence was foundas 4.2% of pSLEpatients [4]. Acute pancreatitis as an initial manifestationof pSLE patients is a rare entity. It can usually occur during active disease states,flaresand remission states [5].

Abdominal pain, raised serum amylase and/or lipase activity, and characteristic imaging findings are essential fordiagnosingacute pancreatitis in children [6]. Acute pancreatitis may be an early manifestation of this disease, occurring most frequently in the first two years of SLEdiagnosis [7].

The etiopathogenesis of pancreatitis in SLE is unknown. Literature suggested that vasculitis, immune-mediated injury, thromboembolic events and generalized serositisare responsible for this event [4,8].The rarity of pSLE and acute pancreatitis make it more challenging for a treating physician to diagnose and manage them. The outcome is variable and often affected by the delay in reaching the diagnosis. This could happen particularly when pancreatitis isa presenting manifestation of SLE [9] .The mortality rate of lupus-related pancreatitis in children is very high [10].

In this report,we present a casewithpSLEwith acute pancreatitisas an initial presentation.This atypical and life-threatening presentation of pSLEencouraged us to report this case so that awareness can be created and early management may be ensured.

Case

A13- year old girl presented withsevere epigastric pain and vomiting for two days. She also hada history of low-gradeirregular fever, rash over the face and extremities and polyarthralgiafor two months.No history of jaundice, diarrhea, constipation, sore throat, dysuria or family history of autoimmune disease were found.For the above complaints, she was managed initially with analgesics, omeprazole and antibiotics. However, she did not respond to these treatments and got admitted in the department of pediatrics,Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka. On admission, she was mildly pale, lethargic,non-icteric and her vitals were within normal limits.Non-specific erythematous rashes were distributed over the face and extremities. Ulcers were present on the hard palate and alopecia were also present. The girl also hadcervical lymphadenopathy.Her abdomen was distended with diffuse tenderness more in the upper regions and bowel sound was intact.The girl had moderate lymphadenopathy.Other systemic examinations were normal.

Laboratory investigations found low haemoglobin (9.9gm/dl),low total count(3500/mm^3)increase neutrophils (2145/$cumm^3$), low lymphocyte (1280/mm^3) and normalplateletcount (200000/mm^3),ESR was 29 mm in the first hour and blood film showed normochromic normocytic anemia. Cultures of blood and urine showed no growth.Her liver enzymes were raised SGPT - 237 U/L, Alkaline phosphatase 75IU/L.Serum bilirubin, serum albumin and coagulation profile (PT, APTT) was normal.Viral markers for hepatitis (HBsAg, Anti-HAV IgM and Anti- HEV IgM) were negative. Serum electrolytes, serum calcium andrandom blood sugar were also normal. She had elevated serum lipase-6124 U/L and serum amylase 473 U/L.

Chest x-ray and plain x-ray of the abdomen showed normal findings. Ultrasonography of the whole abdomen revealed an edematous pancreas and evidence of gallstone and ascites were absent.

The girl was initially treatedwithconservative management includingnothing per oral, intravenous fluid, analgesics and empiricalantibiotics. Her abdominal pain was gradually improved and serum lipase and amylase levels returned to normal by the 14^{th} day of hospital admission. Further laboratory analysis showed negative ANA (IF method), anti-ds-DNA, and anti cardiolipin antibodies, but she had positive anti- smith antibody and low C_3level. Routine urine examination, 24- hour urinary total protein (0.37 gm/day) and serum creatinine (0.3mg/dL) were normal. Lipid profiles were within normal limit and coombs test were also negative. Punch biopsy of skin was performed, which showedhyperkeratosis, hydropic degeneration of basal layer and thick basement membrane of epidermis. Direct immunoifluroscenece (DIF) showed deposition of IgA (1+), IgG(1+), C3(1+) andIgM(trace). These findings were consistent with discoid lupus erythematosus(DLE). Finally, this girl was diagnosed as a case of SLE with acute pancreatitis.

We managed the girl withintravenous methylprednisolone (30mg/kg/day) for three consecutive days, followed by oral prednisolone for 4 weeks and later on tapered with other

supportive management. Finally, hydroxychloroquine (5 mg/kg/day) was added after normalization of liver function.The patient was discharged after three weeks of hospital stay. She is now stable and regularly attending our follow-up clinic and maintaining remission with the above treatment.

Discussion

Acute pancreatitis associated with pSLE is a diagnosis of exclusion that could be confirmed after ruling out the other possible causes. It is an early manifestation of lupus most commonly seen within the first year of diagnosis and often goes parallel with lupus activity. There is no correlation with serology except in severe cases when complements can be low. Disease course is often challenging by local and systemic complications with higher mortality in severe cases. Most commonly used drugs for management included steroids and cyclophosphamide.In children, blunt abdominal trauma, multisystemdisease, biliary stones and drug toxicities are most common etiologies for acute pancreatitis [6]. Our patient presented with acute pancreatitis and alsohad manifestations of pSLE.The association between SLE and pancreatitis was first documented by Reifenstein et al. in 1939 [11]**.** Most pancreatitis cases reported in SLE patients with long-standing disease courses had multi-organ involvement and weretreated with steroids and immunosuppressive therapy [12]. The occurrence of pancreatitis as an initial manifestation of SLE is very infrequent.Up to date literature survey documented 100 cases of SLE-associated pancreatitis and among them,only 12patients had acute pancreatitis as an initial manifestation [13].

The pathological mechanism of acute pancreatitis in SLE is not only complicated but also multifactorial. Vasculitis, microthrombi, interstitial edema, immune complex deposition, occlusion of arteries and arterioles, autoantibody production,abnormal cellular response and drug toxicities are major contributing factorsfor the development of acute pancreatitis in SLE [5,8,14].

The diagnostic criteria of acute pancreatitis require two of the following three criteria: Typical abdominal pain; amylase and/or lipase levels that are three or more times the upper limit of normal; and typical abdominal ultrasoundor an abdominal CT scan or MRI of the pancreas [15].

Acute pancreatitis symptoms may range from self-limiting disease to severe fulminant form. Also, subclinical pancreatitis may occur in SLE patients [16]. Our patient had typical abdominal pain with significantly raised pancreatic enzymes and ultrasonography findings suggestive of acute pancreatitis.

An Indian report showed acute pancreatitis as the initial feature. Later on, the patient developed bilateral pleural effusion, ascites and persistent haematuria, proteinuria and azotemia. All serological tests (ANA, anti-ds-DNA, anti-smooth muscle antibody) were positive and renal biopsy showed grade IV nephritis. After receiving IV methylprednisolone and cyclophosphamide, this patient was stable and discharged from the hospital [12].

Qadiaryet al. in their study in Morocco, observed two adolescent girls presented initially with acute pancreatitis and finally diagnosed as pSLE.One case presented with neurological involvement and another one with joint involvement along with abdominal symptoms. They were treated with IV steroid and cyclophosphamide [14].

In our case oral ulcers, non-specific rashes, alopecia arthralgia and features of acute pancreatitis were present. Among the antibodies, the only anti-smith antibody was positive, along with low C3 level. Skin biopsy resultswere suggestive for discoid lupus erythematosus (DLE). ACR revised classification criteria were not fulfilled, but Systemic Lupus International Collaborating Clinic (SLICC) classification criteria forSLE were fulfilled in this index case [17]. An adolescent girl with multisystem involvement (Musculoskeletal, skin and gastrointestinal) and skin biopsy findings finally guided us to diagnose this girl as SLE with acute pancreatitis. We meticulously searched for the common causes of acute pancreatitis in our patients, namely pancreatic duct obstruction, trauma and medications. Finally, we excluded all these causes and established the diagnosis of lupus pancreatitis in our case.

Current management principlesof acute pancreatitis include pain management, aggressive fluid resuscitation, intensive monitoring and support of organ function [18]. An antibiotic restrictive policy should be adopted. There is no role for prophylactic antibiotics regardless of the severity of acute pancreatitis [19]. All patients should be closely watched for the development of complications including secondary infection. Acute pancreatitis in lupus is usually treated with immunosuppressive and supportive management [20].The treatment of SLE pancreatitis is steroid [21]. Our patient didnot receive steroids previously and evidence showed that steroids do not trigger acute pancreatitis[11].Instead, steroids can significantly improve the outcome of pancreatitis in SLE patients because of the immunosuppressive effect [22]. Recent studies recommendedthe administration of steroids during the acute episodes of SLE pancreatitis [23]. Our patient was treated with glucocorticoids;after that, her clinical symptoms gradually improved,decreasing pancreatic enzymes

and normalizing imaging findings.We also added hydroxychloroquine in our caseafter normalization of liver function.

Prognosis of SLE-related pancreatitis is guarded. Our patient hada complete recovery. The mortality rate of SLE related pancreatitis range from 22%-45% [4,19]. The mortality rate was higher in severe organ involvement (renal, neurological and haematological) in pSLE patients associated with acute pancreatitis[14].In our case, presenting features were suggestive for severe organ involvement. Gastrointestinal manifestation along with skin andjoints were involved here. Among SLE pancreatitis patients, 22% may experience recurrent pancreatitis and 12% may develop pancreatic pseudocyst[19].Our patient had none of these complications till now.

Conclusion

Acute Pancreatitisis a rareand exceptional initial manifestation of pSLE patients. Physicians should always be suspicious of acute pancreatitis, especiallyin those SLE patients who presentwith severe abdominal pain and vomiting. Early diagnosis and treatment with steroidsare mandatory for proper management to improve the outcome in these patients.

References

1. Lin Q, Zhang M, Tang H, Shen Y, Zhu Y, et al. (2020) Acute pancreatitis and macrophage activation syndrome in pediatric systemic lupus erythematosus: case-based review. RheumatolInt 40: 811-9.

2. Rebecca ES, Stacy PA, Laura EC (2016) Systemic Lupus erythematosus. In:Kliegman RM, Stanton BF, Schor NF, Geme JWS & Behrman RE, editors. Nelson Textbook of Paediatrics, 20th ed.Philadelphia:W B Saunders;2016: 1176-7.

3. Richer O, Ulinski T, Lemelle I, Ranchin B, Loirat C, et al. (2007) Abdominal manifestations in childhood-onset systemic lupus erythematosus. Ann Rheum Dis 66: 174-8.

4. Campos L, Omori C, Lotito A, Jesus A (2010) Porta Gand Silva C.Acute pancreatitis in juvenile systemic lupus erythematosus: a manifestation of macrophage activation syndrome. Lupus 19: 1654-8.

5. Wang F (2005) Acute pancreatitis as an initial symptom of systemic lupus erythematosus: A case report and review of the literature. World Journal of Gastroenterology 11: 47-66.

6. Steven LW, Michael W (2016) Acute Pancreatitis.In:Kliegman RM, Stanton BF, Schor NF, Geme JWS & Behrman RE, editors. Nelson Textbook of Paediatrics, 20th ed.Philadelphia:W B Saunders 2016: 1913-4.

7. BreuerG, Baer A, Dahan D, Nesher G (2006) Lupus-associated pancreatitis. Autoimmunity Reviews 5: 314-8.

8. Yang Y, Ye Y, Liang L, Wu T, Zhan Z, et al. (2012) Systemic-Lupus-Erythematosus-Related Acute Pancreatitis: A Cohort from South China. Clinical and Developmental Immunology 2012: 1-8.

9. Makol A, Petri M (2009) pancreatitis in systemic lupus erythematosus: frequency and associayted factors-a review of the Hopkins Lupus Cohort. J Rheumatol 37: 341-5.

10. Tian X (2010) Gastrointestinal involvement in systemic lupus erythematosus: Insight into pathogenesis, diagnosis and treatment. World Journal of Gastroenterology 16: 2971-7.

11. ReifensteinE (1939) A variable symptom complex of undetermined etiology with fatal termination. Archives of Internal Medicine 63: 553-4.

12. Eaker EY, Toskes PP (1989) Systemic lupus erythematosus presenting initially with acute pancreatitis and a review of the literature. Am J Med Sci 297: 38-41.

13. Bandyopadhyay D (2015) Acute pancreatitis- As a Presenting Manifestation of Systemic Lupus Erythematosus. American Journal of Medical Case Rep 3: 155-7.

14. Bairy I, Berwal A, Prabhu M, Budania L (2018) Systemic Lupus Erythematosus with Acute Pancreatitis as an Initial Presentation. Journal of clinical and diagnostic res 12: 1-3.

15. El Qadiry R, Bourrahouat A, Aitsab I, Sbihi M, Mouaffak Y, et al. (2018) Systemic Lupus Erythematosus-Related Pancreatitis in Children: Severe and Lethal Form. Case Reports in Pediatrics 2018: 1-4.

16. Xiufeng L, Baofeng X, Xifan L, Ziwei L, YuanyuanGao RL (2020) A case report and literature review of systemic lupus erythematosus with sustained elevated serum amylase and lipase levels. Int J ClinExp Med 13: 2641-8.

17. Petri M, Orbai AM, Alarcon GS, Gordon C, Merrill JT, Fortin PR et al. Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus, Arthritis Rheum 2012; 64 (8): 2677-86.

18. Banks PA, Conwell DL, Toskes PP (2010) The management of acute and chronic pancreatitis. GastroenterolHepatol (NY) 6: 1-16.

19. Mourad MM, Evans R, Kalidindi V, Navaratnam R, Dvrokin L,Branhall SR (2017) prophylactic antibiotics in acute pancreatitis: endless debate.Ann R Coll Sung Engl 99: 107-12.

20. Nesher G, Breuer GS, Temprano K (2006) Lupus associated pancreatitis. Semin. Arthritis Rheum 35: 260-7.

21. Duncan H, Achara G (2003) A Rare Initial Manifestation of Systemic Lupus Erythematosus--Acute Pancreatitis: Case Report and Review of the Literature. The Journal of the American Board of Family Medicine 16: 334-8.

22. Jia Y, Ortiz A, MccallumR, Salameh H, Serrato P (2014) Acute Pancreatitis as the Initial Presentation of Systematic Lupus Erythematosus. Case Reports in Gastrointestinal Medicine 2014; 1-4.

23. Pascual RV, Duarte RA, Villa AR, Hernández CB, Alarcón SD, et al. (2004) Systemic lupus erythematosus as a cause and prognostic factor of acute pancreatitis. J Rheumatol 31: 707-12.