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Case Report

Acute pancreatitis: An unusual presentation of systemic lupus erythematosus

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ABSTRACT

Systemic lupus erythematosus (SLE) is a multi-system and chronic inflammatory autoimmune disease characterised by the presence of autoantibodies and the creation of immune complexes, coming up with protean manifestations. Its occurrence in children is rare and acute pancreatitis as the presenting manifestation of childhood SLE is exceptional. Diagnosis of lupus pancreatitis is clinical, biological and radiological. Here, we report an 8 year 8-month-old young girl who presented to our hospital with abdominal pain, nausea and vomiting. She also had a fever, alopecia, oral ulceration and glossitis. Her investigations showed elevated serum amylase and lipase levels, suggestive of acute pancreatitis. Other investigations revealed pancytopenia, low complement, microalbuminuria and high antinuclear antibodies level, indicative of SLE. As a result, the diagnosis of lupus pancreatitis was retained. Her clinical symptoms and signs markedly improved after a course of glucocorticoid and immunosuppressive therapy. The child is now being brought for regular follow-up and is doing well.

Keywords: Systemic lupus erythematosus, Paediatric systemic lupus erythematosus, Lupus pancreatitis

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune inflammatory disorder with multi-system involvement, potentially involving almost every system and organ in the body.[1] SLE has several phenotypes and varying clinical presentations ranging from mild mucocutaneous manifestations to multi-organ and severe central nervous system involvement. Although gastrointestinal symptoms are frequent constituting 19% in paediatric SLE and 8–40% in adult SLE, pancreatitis is an extremely rare manifestation of SLE, comprising 6% and 3–8% in children and adults, respectively.[2] Thus, acute pancreatitis is more severe, occurs more frequently and is associated with higher mortality in paediatric SLE patients when compared to adults with SLE.[3] It has life-threatening severity despite well-conducted treatment. The occurrence of SLE and pancreatitis are uncommon in children and the comorbidity of these two conditions is quite rare. Pancreatitis usually occurs during an active disease state, flares and remission. It is infrequent for acute pancreatitis to be the presenting manifestation of SLE.[3,4] The exact pathogenesis is not well understood, but literature attributes it to vascular complications (such as vasculitis, non-inflammatory vasculopathy, thrombosis related to anti-phospholipid antibodies) and toxic-metabolic causes (treatment with certain drugs such as steroids).[5] Lupus pancreatitis can be confirmed when a child with active SLE presents with two out of three of the following: Clinically typical acute abdomen, >3-fold increase in serum amylase/lipase and evidence from imaging findings. Although rare, the mortality rate is very high, ranging up to 45% and 22%

of patients with lupus pancreatitis may have recurrent pancreatitis attacks, and 12% of patients develop pancreatic pseudocysts.[6]

CASE

An 8 year 8-month-old female child, 2nd in birth order, born to a non-consanguineously married couple, developed and immunised for age, was hospitalised in paediatric intensive care unit. The child's mother gave a history of fever for 3 months, low-grade, intermittent in nature, associated with abdominal pain for 2½ months, stabbing in nature, exaggerated after having food and was associated with nausea and vomiting. She also had a history of poor appetite, weight loss and contact with a tuberculosis patient. She otherwise had no medical history or family history of autoimmune disease.

On examination, the child was emaciated, had patchy hair loss [Figure 1], looked pale and had oral ulcers with glossitis. On deep palpation, epigastric and periumbilical region tenderness with hepatosplenomegaly was noted. She was otherwise afebrile with stable vital signs and the rest of the somatic examination, including the neurological examination, was ordinary.

Biologically, outside/previously done blood investigations serially showed leukopenia, thrombocytopenia, elevated liver enzymes and raised ferritin. Ultrasonography (USG) abdomen and pelvis showed mild hepatomegaly and mesenteric lymphadenopathy with minimal interloop fluid.

At admission, pancytopenia was observed at 8.2 g/dL without signs of haemolysis, thrombocytopenia at 60,000/µL and lymphopenia at 3900/µL (N/L/M - 46/40/5), Erythrocyte sedimentation rate (ESR) at 54 mm at the 1st h, negative C-reactive protein, C3 levels sent were 28 (normal range: 90–180 mg/dL) and there was microalbuminuria with normal renal function. Aspartate transaminase and Alanine transaminase were 575 U/L and 155U/L, respectively. The diagnosis of pancreatitis was strongly suspected and confirmed by hyperamylasaemia and hyperlipasaemia – 1197 IU/L (normal <220 U/L) and 9449 IU/L (normal <190 U/L), respectively.

The child was further worked up for tuberculosis and was reported negative. Blood samples sent for culture and sensitivity were sterile. Widal, Weil-Felix, malarial parasite tests and HIV serology were also negative. Bone marrow aspiration/cytology showed dimorphic erythropoiesis with mild dyspoeisis and myeloid series showed left shift with toxic changes. Bone marrow biopsy showed megaloblastic erythropoiesis. The diagnosis of SLE was retained in front of the multisystemic symptoms and met the criteria of the American College of Rheumatology; the nucleosomes were positively associated with C3 hypocomplementemia. Based on clinical and laboratory criteria, it was concluded to be SLE with pancreatitis with pancytopenia. Once the clinical diagnosis was obtained, methylprednisolone pulse therapy was administered, followed by oral prednisone and immunosuppressive therapy with Mycophenolate mofetil and other required supportive measures.

Hydroxychloroquine was added after liver function normalised. She responded well to the therapy; her clinical symptoms and signs subsided eventually and pancreatic enzymes gradually normalised. The aetiology of acute pancreatitis was to be investigated with imaging, but the child's parents could not afford to pay for the same. The high titre of antinuclear antibodies and low level of complement components during the pancreatitis attack suggested that pancreatitis may have been due to SLE exacerbation and not related to drug therapy. The child was discharged on oral steroids after 3 weeks of hospital stay. She attends our follow-up clinic regularly and is now stable with a good general condition for the past 6 months, routine blood tests and urine analyses and maintaining remission with the follow-up care for SLE.

DISCUSSION

SLE, an autoimmune disease involving multiple systems with protean clinical manifestation, is uncommon in the paediatric age group. Despite gastrointestinal involvement being a prevalent situation, pancreatitis is rare in both adult and paediatric SLE, but when it occurs, it is aggressive in children and is linked to high mortality if not diagnosed and treated promptly. Association between SLE and pancreatitis was first propagated by Reifenstein and Reifenstein in 1939.[7] Pathogenesis, as described earlier, is not clear, but studies have attributed it to vascular complications (such as vasculitis), deposition of immune complexes associated with occlusion of arteries and arterioles and interstitial oedema.[8]

Other hypotheses such as the production of autoantibodies, the abnormal immune response of cells of the pancreas and toxic-metabolic causes (due to drugs, alcohol intake) have also been put forward.[9]

Diagnosis of SLE-related gastroenteropathies is usually based on two out of three among typical abdominal pain, lab results of abnormal pancreatic enzymes and ultrasound/magnetic resonance/tomographic signs in favour.[10] Cases of SLE having symptoms such as pain abdomen and vomiting are treated with proton pump inhibitors, mistaken for gastritis/side effects of multiple therapies and pancreatic enzymes do not get measured. The rarity of lupus pancreatitis makes it challenging for the treating physician to diagnose and manage. Pancreatic enzymes and imaging are to be performed in all the patients who develop abdominal pain as it is an avoidable cause of pancreatitis. Few children fulfilled guidelines for macrophage activation syndrome, this association explaining that pancreas could be target organ of this syndrome.[11] Diagnosis should only be made after

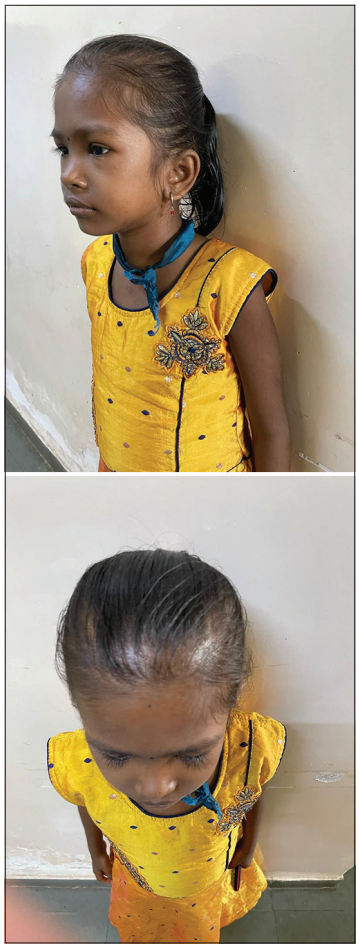

Figure 1: Alopecia areata (patchy hair loss) in this child with SLE.

excluding pancreatic duct obstruction, trauma and toxic-metabolic and mechanical aetiologies of pancreatitis such as gall stones, medication and hypertriglyceridemia.

In immunocompromised children, viral causes of HIV are included in the study.[9] In children with insidious onset of abdominal pain, non-invasive investigations such as USG and abdominal computed tomography (CT) should be considered. CT/MRI may identify abdominal abscesses, pancreatic pseudocysts and organomegaly. Abdominal USG can show bowel wall thickening. Invasive investigations such as Barium studies, gastroscopy, colonoscopy (with biopsy) and specialised techniques such as gallium and indium-111 white cell scanning can help differentiate and diagnose.[12]

Management of lupus pancreatitis has been controversial as corticosteroids have been proposed as a possible cause of acute pancreatitis in the past.[13] In contrast, in most recent studies, as well as in the current series, there are reports of pancreatitis being the initial presentation of SLE and also several cases with pancreatitis occurring in the absence or withdrawal of corticosteroids and also initiating increasing steroid treatment results in resolution of pancreatitis in most of the patients.[2,9,14] Thus, the therapeutic benefit seems to be far above the risk of exacerbation of pancreatic lesions.

Many children with SLE complaining of pain abdomen may be diagnosed with gastritis or other conditions such as ascites, sepsis or treatment-induced events (in children who received drugs with potential gastrointestinal side effects, in the absence of any other cause), pancreatic enzymes are not measured.[15] Mild pancreatitis can be subclinical or resolve spontaneously, or due to ongoing corticosteroids course for active SLE, at least some of lupus pancreatitis cases are not diagnosed. Therefore, pancreatitis in SLE might be underestimated.[2]

Campos *et al.*[16] showed that renal and joint involvement is usually seen in Lupus pancreatitis. Still, our child has not had any such effect, which could have also contributed to the excellent prognosis.[17] Early diagnosis, suspected in front of any abdominal pain and appropriate treatment with high-dose corticosteroids and immunosuppressive therapy has shown improvement in prognosis. According to International Study Group of Pediatric Pancreatitis: In search for a cure (INSPPIRE) standardised definitions, pancreatitis in SLE patients is mostly an acute subtype with rare recurrence or progression to chronic damage.[18] Rarely, in severe cases, therapeutic plasma exchange and intravenous gamma-globulin infusion may be beneficial.

CONCLUSION

Acute abdomen in SLE patients is easily mistaken for gastritis and proton-pump inhibitors and analgesics are usually tried. Physicians should always have a high index of suspicion of pancreatitis in such cases and immediately work up for pancreatic enzymes and/or imaging. Since lupus pancreatitis has a high mortality, it needs to be diagnosed early as aggressive treatment may be life-saving.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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