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Case Report

The Acute Pancreatitis of SLE: An Unusual Presentation

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Abstract

pediatric SLE, as it seldom occurs in children. Lupus pancreatitis can be diagnosed by radiological, biochemical, and clinical means. Here, we are reporting the case of an 8-year-old girl who came to our hospital complaining of nausea, vomiting, and abdominal pain. She also experienced glossitis, mouth ulcers, baldness, and a fever. Her tests revealed increased lipase and amylase levels in the serum, which may indicate acute pancreatitis. Additional tests found SLE-related pancytopenia, poor complement, microalbuminuria, and elevated ANA levels. Consequently, the diagnosis of pancreatitis caused by Lupus was kept. She had glucocorticoid and immunosuppressive medication, and following that, her clinical symptoms and indicators significantly improved. The child is doing well and is currently being brought back for routine follow-up.

Keywords: Systemic lupus erythematosus (SLE) ~ Paediatric SLE (pSLE) ~ Lupus pancreatitis

Introduction

An autoimmune inflammatory disease that can affect nearly every system and organ in the body is called systemic lupus erythematosus (SLE) (1). SLE exhibits a number of phenotypes and differs in its clinical manifestations, from minor mucocutaneous symptoms to severe CNS involvement involving several organs. Despite being common—consisting of 8–40% in adult SLE and 19% in pediatric SLE—gastrointestinal symptoms are rare. A very uncommon symptom of systemic lupus erythematosus (SLE), pancreatitis affects 6% and 3-8% of children and adults, respectively (2) Therefore, compared to adults with SLE, patients with pSLE experience acute pancreatitis more frequently, with a higher degree of severity, and with a higher fatality rate (3). Even with effective therapy, the intensity is life-threatening. SLE and pancreatitis are uncommon in children, and it is quite uncommon for these two illnesses to coexist. Pancreatitis typically develops during an active phase of the illness, with flare-ups and remissions. Acute pancreatitis is not usually the first symptom of SLE to appear (3,4). Although the precise etiology is unknown, research points to toxic-metabolic factors (drug usage such as steroids) and vascular consequences (vasculitis, non-inflammatory vasculopathy, thrombosis associated with anti-phospholipid antibodies) (5). The diagnosis of lupus pancreatitis can be made when a child with active SLE has two or more of the following three symptoms: Clinical signs include a characteristic acute abdomen, a serum lipase and amylase rise of more than three

times, and imaging results supporting the diagnosis. Even though lupus pancreatitis is uncommon, the death rate is very high, reaching up to 45%. Additionally, 12% of patients get pancreatic pseudocysts, and 22% of patients may experience recurring pancreatitis attacks (6)

Case

8 years 8 months old, female, born into a non-consanguineous marriage, ranked second in birth order, matured and immunized for her age, and was admitted in paediatric general ward. The mother of the kid reported that the child had experienced low-grade, sporadic fever for three months, which was linked to intermittent abdominal pain that had become accentuated after eating and was accompanied by nausea and vomiting since two and a half months. In addition, she had previously experienced weight loss, decreased appetite, and interaction with a tuberculosis patient. Other than that, she had no family history of autoimmune disease or medical history.

Examining the child revealed that he was malnourished, had patchy hair loss, was pale, and developed glossitis and mouth ulcers. Deep palpation revealed hepatosplenomegaly together with discomfort in the periumbilical area and epigastrium. The rest of the somatic examination, including the neurological examination, was routine, and she remained afebrile with stable vital signs.

From a biological perspective, past blood studies conducted outside the body have consistently demonstrated increased ferritin, thrombocytopenia, leukopenia, and liver enzymes. The USG revealed mesenteric lymphadenopathy with little interloop fluid and mild hepatomegaly in the belly and pelvis.

Pancytopenia (8.2 g/dL) without hemolysis signs was noted upon admission. Thrombocytopenia (60,000/µL) and lymphopenia (3900/µL) (N/L/M - 46/40/5) were also noted. The first hour's ESR was 54 mm, CRP was negative, C3 levels were 28 (normal range: 90–180 mg/dl), and microalbuminuria was present with normal renal function. There were 575 U/L for AST and 155 U/L for ALT. Strong suspicions of pancreatitis were supported by hyperamylasemia and hyperlipasemia, both of which were 1197 IU/L (normal <220).

The youngster had additional testing for tuberculosis, but the results were negative. Sterile blood samples were supplied for sensitivity testing and culture. HIV serology, malarial parasite testing, Weil-Felix, and Widal tests were all negative. Aspiration and cytology of the bone marrow revealed dimorphic erythropoiesis with mild dyspoeisis, and the myeloid series revealed a left shift with toxic alterations. Macroloblastic erythropoiesis was revealed by a bone marrow biopsy. The American College of Rheumatology (ACR)

criteria were met, and the diagnosis of Systemic Lupus Erythematosus (SLE) was maintained in light of the multisystemic symptoms; nucleosomes were positively correlated with C3 hypocomplementemia. SLE with pancreatitis and pancytopenia was determined based on clinical and laboratory criteria. Following the clinical diagnosis, immunosuppressive medication and oral prednisone were given, then methylprednisolone pulse therapy.

Following the return of normal liver function, hydroxychloroquine was introduced. Her clinical symptoms and indicators subsequently diminished, and her pancreatic enzymes gradually returned to normal, indicating that she reacted effectively to the therapy. Imaging was to be used to look into the cause of the acute pancreatitis, but the child's parents were unable to pay for it. A pancreatitis episode may have been caused by a worsening of SLE rather than a reaction to medication therapy, as indicated by the high titer of antinuclear antibodies and low level of complement components during the attack. After three weeks in the hospital, the child was released on oral steroids. She has been stable for the previous six months, undergoing routine blood tests and urine analysis, maintaining remission with the follow-up, and consistently attending our follow-up clinic.

Discussion

In the pediatric age range, systemic lupus erythematosus (SLE), an autoimmune disease affecting several systems with erratic clinical expression, is rare. Pancreatitis is uncommon in both adult and pediatric SLE, although gastrointestinal involvement is common. When it does occur, it is aggressive in children and has a high fatality rate if left untreated. Reifenstein et al. originally reported a link between pancreatitis and SLE in 1939 (7). As previously mentioned, the pathogenesis of this condition is unclear; nevertheless, research has linked it to interstitial oedema, immunological complex deposition linked to blockage of arteries and arterioles, and vascular consequences such vasculitis (8).

Additional theories include the development of autoantibodies, aberrant immunological responses in pancreatic cells, and toxic-metabolic reasons (caused by drugs, alcohol intake etc) (9).

The diagnosis of gastroenteropathies related to SLE is typically made on the basis of two out of three factors: normal abdominal discomfort, abnormal pancreatic enzyme test values, and symptoms that are favorable on ultrasonography, magnetic resonance imaging, or tomography (10). Proton pump inhibitors are used for SLE cases exhibiting symptoms such as abdominal discomfort, vomiting, and side effects from several

medications. Pancreatic enzyme levels are not assessed in these cases. The treating physician finds it difficult to detect and treat lupus pancreatitis due to its rarity. Since pancreatitis can be prevented, all individuals experiencing abdominal pain should have pancreatic enzyme testing and imaging done. The pancreas may be the target organ of macrophage activation syndrome, as shown by the fact that few children met its standards (11). Only after ruling out pancreatic duct obstruction, trauma, and toxic-metabolic and mechanical causes of pancreatitis, such as gallstones, drugs, and hypertriglyceridemia, can a diagnosis be determined. HIV infection is caused by viruses in youngsters with weak immune systems (9).

Non-invasive tests like abdominal computed tomography (CT) and ultrasonography (USG) should be taken into consideration for youngsters whose abdomen pain develops slowly. Organomegaly, pancreatic pseudocysts, and abdominal abscesses can all be detected by CT/MRI. Bowel wall thickening might be seen on an abdominal USG. invasive tests such as colonoscopies (with biopsy), gastroscopies, and barium studies and specialized methods such as white cell scanning with gallium and indium-111 can aid in diagnosis and differentiation (12).

Since corticosteroids have previously been suggested as a potential cause of acute pancreatitis, the management of lupus pancreatitis has been controversial (13). On the other hand, pancreatitis has been reported as the first symptom of SLE in the majority of recent studies and the current series. There have also been several cases of pancreatitis that occur when corticosteroids are stopped or not taken at all, and starting to increase the dosage of steroids cures pancreatitis in the majority of patients (2,9,14). Therefore, the chance of the pancreatic lesions getting worse appears to be greatly outweighed by the therapeutic benefit.

When a child with SLE complains of stomach discomfort, pancreatic enzymes aren't checked, and the diagnosis may be made of gastritis, ascites, sepsis, or treatment-induced events (in children who got medications with potential GI side effects, in the absence of any other explanation) (15). At least some cases of lupus pancreatitis go undiagnosed because of continuing corticosteroid treatment for active SLE, or because mild pancreatitis is subclinical or resolves spontaneously. Pancreatitis in SLE may therefore be underdiagnosed (2).

According to Campos et al. (16), renal and joint dysfunction is typical in pancreatitis caused by lupus. However, our youngster hasn't experienced any of these effects, which may have also added to the favorable prognosis (17). The prognosis has improved with early detection, suspected before any stomach pain, and adequate treatment with immunosuppressive medication and high-dose corticosteroids. The majority of pancreatitis in SLE patients is an acute subtype with a rare recurrence or progression to chronic damage,

according to INSPPIRE standard classifications (18). Intravenous gamma-globulin infusion and therapeutic plasma exchange may be helpful in rare, severe situations.

Conclusion

Since gastritis and acute abdomen are frequently confused in SLE patients, analgesics and proton-pump inhibitors are typically used. In such circumstances, doctors should always have a high index of suspicion for pancreatitis and should get pancreatic enzymes and/or imaging as soon as possible. Given the significant fatality rate of lupus pancreatitis, Early diagnosis is necessary since intensive treatment could save lives.

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