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CASE STUDY

ACUTE PANCREATITIS IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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ABSTRACT

Acute pancreatitis is a rare entity when presenting in a patient with Systemic Lupus Erythematosus. Since it was first described in 1939 less than 200 cases have been reported in literature. We are reporting a case of a 17 year old girl, a diagnosed case of SLE, who had developed sudden onset abdominal pain, and subsequently diagnosed with Lupus induced Acute Pancreatitis. She was started on steroid therapy after the diagnosis but consequently succumbed to the illness, after developing lupus pancreatitis related ARDS. This case report thus intends to highlight the need for early diagnosis with a raised index of suspicion with pancreatitis being the possible diagnosis in SLE patients presenting with pain abdomen and the need for early intervention in such patients.

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INTRODUCTION

Systemic Lupus Erythematousus (SLE) is a multi-systemic autoimmune connective tissue disease. Gastrointestinal symptoms are common in SLE patients, in about 19.2%–50% patients (Yang *et al*., 2012), but though they may not be as common as lupus nephritis, they are clinically important as they can be life threatening if not treated promptly. (Tian and Zhang, 2010) Acute pancreatitis is an unusual and rare manifestation of SLE.Since it was first described in 1939 by Reifenstein *et al*, less than 200 cases have been reported in literature[17]. It may present as an initial presentation of SLE, or more commonly, after the diagnosis of SLE, with patients complaining of acute onset abdominal pain.Current literature suggests that sudden withdrawal of low dose steroids or lack of steroid therapy may be one of the causative factors. (Goel, 2012) One should always keep in mind the diagnosis of acute pancreatitis as a differential diagnosis of pain abdomen in patients of SLE, complaining of pain abdomen, as early diagnosis and prompt treatment with corticosteroids help in decreasing the mortality and morbidity in patients, especially after ruling out the common causes of pancreatitis, such as gall stone, alcohol and medications.

Case report

We report a 17 year old girl, of lean built, a diagnosed case of SLE since June 2015, who had come to our district general hospital with complaints of painful lesions on her hands since 2-3 months, with increase in severity since the past 5-6 days, pain abdomen and joint pain on and off. On examination, the lesions were painful, reddish, and present on her palms and digits, associated with peri-lesional swelling without bluish discoloration of hands and digits on cold exposure. She also complained of few faintly pink coloured lesions on upper limb, back, face, and eyelids that were itchy and worsened on exposure to sunlight. There were tender purpurae present with surrounding blanchableerythma, with few scales, over the palms. She also complained of joint pain, especially in the joints of the hand, but with no limitation of movements, along with easy fatigability and hair loss. She had no complaints regarding menses. Three days after her admission, she complained of sudden onset acute abdominal pain which radiated to the back and vomiting since morning. On examination, patient had a temperature of 38.3 degree Celsius; pulse rate of 104/min, and her blood pressure was 100/60 mm Hg. On per abdomen examination, patient had tenderness in epigastrium withlocalised guarding. Bowel sounds were present in all four quadrants but sluggish in frequency. Examination of the chest revealed a bilateral equal air entry and clear breath sounds. Chest X-Ray showed no abnormalities Routine blood investigations were done, with the CBC showing ahemoglobin count of 9.1gm/dL, white Blood Cell count of 17.65 cell/mm^3, and a platelet count of 145,000 cells /mm^3 . Her renal function tests, liver function tests and serum electrolyte values, serum calcium and serum phosphate values were within normal range. Serum amylase level at 974 U/L and her serum lipase levels were 7598 U/L.Her C-reactive protein counts were raised to the level of >10.5mg/L, and ANA titre was strongly positive with a value of 6.89. Her U1-

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nRNP/Sm antigen test and SS-A and Ro-52 antigen tests also were positive.

An ultrasonography of the abdomen and pelvis was done next, which showed a heterogenous and bulky pancreas, with head measuring 2.4cm and tail measuring 3.2cm, with surrounding peri-pancreatic fluid collection and echogenic surrounding fat, along with mild to moderate ascites. A computed tomography (CT) scan of the abdomen and pelvis was done, which showed a bulky pancreas, appearing heterogenous with few non enhancing areas within its parenchyma (more than 30%) suggestive of acute necrotising pancreatitis with modified CT severity index of 10/10, with extensive peri-pancreatic fat stranding and fluid collection, with few enlarged perigastric group and mesenteric lymph nodes detected, largest measuring 1 X 0.7 mm, with moderate ascites and bilateral mild pleural effusion. (Figures 1,2,3,4)

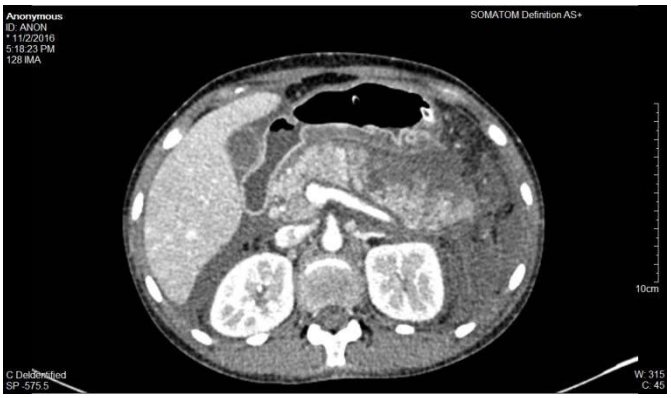

Figure 1. Axial view showing bulky pancreas with necrosis in body and tail and peripancreatic fat stranding. Splenic vein seen and shows absence of filling defect

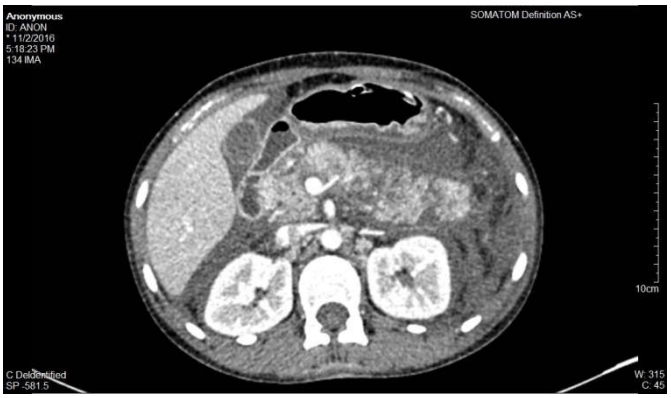

Figure 2. Axial sections showing bulky necrotic pancreas with absence of gall stones and normal pancreatic duct

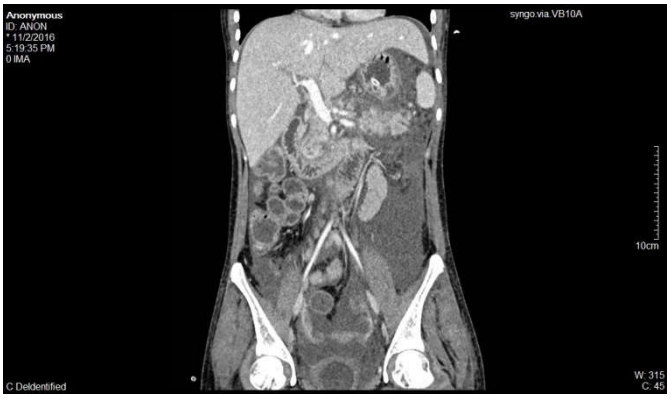

Figure 3. Coronal section showing necrosis involving body and tail of pancreas, with peripancreatic collection, fat stranding and bilateral pleural effusion

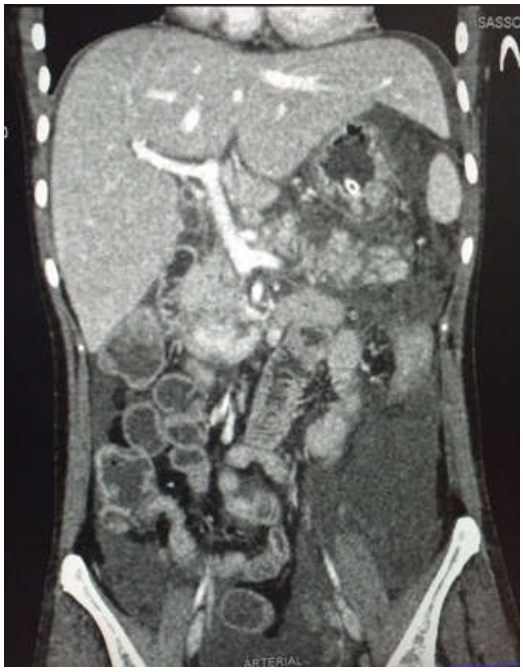

Figure 4. Coronal section showing bulky necrotic pancreas with free fluid in abdomen

A diagnosis of SLE induced pancreatitis was thus considered as the primary diagnosis, as the patient gave no history of alcohol consumption, and intake of medication except for low dose prednisolone. She was started on low dose tablet prednisolone after her previous admission, which was started at 30 mg per day dose and tapered to 10mg per day. The parents did give a history of voluntarily stopping the tablet medication around 1 month prior to the current admission. The patient also had no signs of gall stones on radiological investigations and tested negative for HIV and other STD's. Her Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) score was 11, indicating moderate to severe flare. The patient was managed conservatively with injectable antibiotics, intravenous fluids and started on Injectable Octreotide 100 microgram twice a day and Injectable methylprednisolone 100 mg twice a day. Patient had to be transferred to the Intensive Care Unit, in view of tachypnoea and decreased blood oxygen saturation and pO2 of 52.5mmHg and metabolic acidosis with pH of 6.926 on arterial blood gas analysis, indicating impending respiratory failure.Chest x ray showed patchy consolidation, and she was electively intubated. Inspite of aggressive therapy with steroid and maintaining hydration through a central venous line, her serum Amylase levels continued to rise at a level of 912 U/Land withpersistant sepsis with WBC counts at 15.90 cells/^{mm3}. Patient consequently passed away from a cardiorespiratory arrest possible secondary to ARDS in a setting of lupus induced acute necrotizing pancreatitis. A post mortem could not be done, as the parents did not give consent to it.

DISCUSSION

Systemic Lupus Erythematosus (SLE) is a multi-systemic autoimmune inflammatory connective tissue disease which has several different protean clinical manifestations, with an annual incidence of about 5 cases per 100000 inhabitants and a prevalence of around 52 cases per 100000 inhabitants (Tian and Zhang, 2010; Yuan *et al*., 2013). Acute pancreatitis in a case of SLE is an uncommon, rare, but life threatening complication, the diagnosis of which can be made only after

excluding other common causes of acute pancreatitis, the two most common being gallstones and alcohol toxicity, followed by medications. (Wang *et al*., 2005) Pancreatitis in SLE is an uncommon occurrence. Based on review of literature, the annual incidence of SLE related pancreatitis is estimated to be around 0.4 to 1.1 per 1000 SLE patients (Breuer *et al*., 2006). In a study of SLE patients with pancreatitis, it was seen that acute pancreatitis was more prevalent (0.8%) than chronic pancreatitis (0.1%), and it presented mostly during the active phase of SLE and affected more organs (Wang *et al*., 2016). SLE associated pancreatitis is more frequent in women (88%) with a mean presenting age of 27 years. It can appear as the initial manifestation of SLE in around 22% patients, or within 2 years of the diagnosis of the SLE (in 60%) (Tian and Zhang, 2010; Favarato, 2015). Patients with acute pancreatitis had a higher systemic lupus erythematosus disease activity index (SLEDAI) and higher mortality rates (60%), compared to those SLE patients without pancreatitis (Yang *et al*., 2012; Favarato, 2015; Makol and Petri, 2010). The most common and most characteristic manifestation of SLE induced pancreatitis is abdominal pain, seen in around 80% of patients, with 23% of them complaining of pain radiating to the back (Breuer *et al*., 2006). Patients usually present with involvement of other organ systems, the most common being skin (46%), joints (43%), kidneys (35%), hematological (24%), CNS (21%), cardiac (9%) and pulmonary (8%). (1,6,7,11–13)

The pathogenesis behind SLE pancreatitis is still unclear. Vascular insult is implicated as the main factor. Severe hypertension leading to occlusion of medium and small sized vessels by thrombus, anti-phospholipid syndrome, necrotizing vasculitis, compliment activation and deposition of immune complexes in the arterial wall of the pancreas, have been postulated as the possible mechanisms.An autoimmune reaction involving abnormal cellular immune response or antibody reaction, rather than vasculitis, may also be responsible for the inflammatory reaction. (Lariño-Noia *et al*., 2009) It has been suggested that patients with SLE associated pancreatitis have a higher rate of complications (57%) and higher rates of mortality (up to 45%), especially if untreated, as compared to SLE patients without pancreatitis and in non-SLE patients with pancreatitis. The complications that are seen can be respiratory failure (22%), ascites (19%), pleural effusion (18%) acute renal failure (14%) and circulatory shock (12%). (3,6,7,15) Treatment can be started with azathioprene and glucocorticoids, after ruling out common causes of pancreatitis, such as hepatobiliary pathology, alcohol induced trauma, hypercalcemia, hypertriglyceridemia or medications. (Rodriguez *et al*., 2014) These medications decrease mortality and improve overall prognosis, and even though there has been a concern about these two medications causing pancreatitis on their own, there seems to be no firm evidence confirming this. Also, patients taking steroid therapy prior to onset of pancreatitis had a better prognosis compared to those who were not started on steroids. It was also seen that lack of or withdrawal of prior steroid usage was significantly associated with pancreatitis, especially withdrawal of lose dose maintenance steroid therapy, which was associated with severe pancreatitis compared to mild pancreatitis. (Goel, 2012; Breuer *et al*., 2006; Pascual-Ramos *et al*., 2004; Wang *et al*., 2011)

Conclusion

SLE induced pancreatitis is a clinically challenging diagnosis, and one that should be kept in one's armamentarium of knowledge as a possible cause of abdominal pain in SLE patients, along with differential diagnoses like lupus mesenteric vasculitis, protein losing enteropathy, intestinal pseudo-obstruction and irritable bowel disease. The patient discussed above was a diagnosed case of SLE, who had developed acute pain abdomen, with a history of prior tapering and stoppage of steroid medication. She also had complaints of skin lesions and joint pains. Subsequently, she developed complications of pancreatitis, including pleural effusion and ARDS, and passed away. These facts, aided by a review of the literature discussed above, helped us in the diagnosis of SLE associated Acute Pancreatitis. Thus a need for high vigilance and a keen eye is highly important, for diagnosing this rare entity. Further research and a call for a more meticulous reporting of each case of SLE pancreatitis is required, so as to aid in better understanding of the disease itself as a whole, and to develop a strategy for management of this disease, which has such high morbidity and mortality rate.

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