

Case Report of Pancreatic Pseudocyst in a Patient with Systemic Lupus Erythematosus

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Citation

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Abstract

Introduction: Systemic lupus erythematosus (SLE) is a multisystemic autoimmune disorder that has a wide range of clinical manifestations. Pancreatitis and pancreatic pseudocysts are known complications associated with the disease or attributed to the drugs used in treating SLE. Diagnosis and management of this complication poses a great challenge as SLE patients complaining of abdominal pain can easily be masked by known SLE features of systemic generalized pains and the majority of these patients are on steroids which further masks the symptoms and signs. In our literature search, there has been no reported case of SLE and pancreatic pseudocysts here in the U.K. **Case Presentation:** We present this interesting case report in a 59 year old gentleman with known SLE on steroids and immunosuppressant's presented with symptoms of recurrent abdominal pain and vomiting for 4- 5 months and on screening blood tests suggested pancreatitis and US Scan confirmed pancreatic pseudocyst. He was optimized for surgery and had a Roux-en-Y cystojejunostomy and subsequently discharged from Hospital. **Conclusion:** Pancreatitis as a possibility should be considered in SLE patients presenting with abdominal pains as delay in diagnosing pancreatitis can have grave consequences. The Surgical management of these patients' poses a challenge as they are immunocompromised, undernourished and coagulation is affected and these are further aggravated by steroids and immunosuppressant medications.

Hospital where the work was done: Princess Royal Hospital, Telford, United Kingdom.

INTRODUCTION

Acute pancreatitis is a well known but rare manifestation of systemic lupus erythematosus (SLE) which could lead to the development of pancreatic pseudocysts. Surprisingly, the development of pancreatitis is not usually related to a generalized flare of SLE₁. While a school of thought suspects it is due to medications administered in the treatment of SLE, particularly corticosteroids₂ and azathioprine_{3,4}, another associates it with the widespread vasculitis and thrombosis which is known to occur in the disease. Recent case series reviews have suggested acute pancreatitis as being a manifestation of SLE and not corticosteroids_{5,6} and some studies have actually stressed the importance of steroid therapy in managing SLE associated pancreatitis₆.

Abdominal pain is the commonest symptom in lupus related pancreatitis (88% of patients) followed by nausea and vomiting₅ but as many SLE patients present with

musculoskeletal pain and abdominal symptoms a high index of suspicion will be needed in diagnosing SLE associated pancreatitis.

CASE REPORT

A 45-year-old male non-drinker was diagnosed as having severe SLE in July 2007 with fleeting polyarthritis, urticaria, persistent rash, fever and depressive episodes. Serologically, he was positive for Smith Ag, RNP and Ro antibodies. DNA antibodies were negative. C3 was low at 0.51 and C4 was 0.08. MRI confirmed cerebral lupus. He was initially placed on hydroquinolone, methotrexate and prednisolone which were changed to cyclophosphamide and prednisolone four months later as his lupus was not controlled with the initial regime.

He developed recurrent intermittent abdominal pain, vomiting and left-sided abdominal swelling for 5 months. His blood test showed raised amylase, he had an ultrasound scan and CT scan which confirmed a large pancreatic pseudocyst arising from the body and tail of the pancreas measuring 14.7cm in the largest diameter. There was no

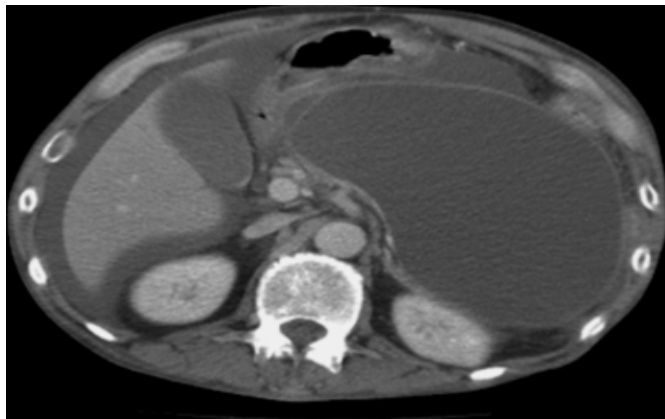
normal tissue in the body or tail of the pancreas but the head appeared normal. There were no gall stones and the biliary tract was normal.

He was referred to our clinic and found to be undernourished and dehydrated. Abdominal examination revealed a large palpable mass 12cm below the left costal margin, which was firm, smooth-surfaced and tender.

He was leucopenic and his ABG, U&Es were normal. He had an elevated GGT of 167U/L, LDH of 704U/L, Amylase of 221U/L, CRP of 77mg/L and an INR of 1.8. His Ranson's score was 1 on admission. A repeat CT scan showed the pseudo-cyst had enlarged to 19.1cm in its widest diameter and there was only a small amount of enhancing tissue in the head and neck of the pancreas (Fig. 1.1).

Figure 1

Figure 1.1



While on admission, he was persistently vomiting and was not tolerating food or fluids. Laparotomy was planned instead of laparoscopy as the cyst size was large and he was optimised for theatre. He underwent Roux-en-Y cystojejunostomy (Fig. 1.2) through a small transverse abdominal incision. Postoperatively, he tolerated oral fluids and food. His abdominal pain settled and he was discharged from hospital on the 8th post-operative day. Subsequently, at two months after the surgery, the patient's symptoms had improved and he had put on weight.

DISCUSSION

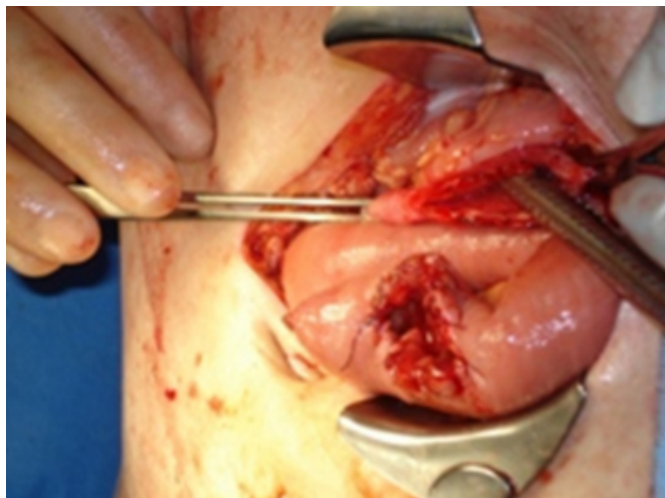
The management of pancreatic pseudocysts in SLE patients poses a challenge to surgical teams in the pre-, intra- and post-operative periods with respect to immunosuppression, coagulopathy, malnourishment, activation of the disease and recovery from surgery. SLE-associated pancreatitis has a higher mortality rate than in non-SLE patients⁵.

Their management should be by a multidisciplinary approach which should include rheumatologists, gastroenterologists, haematologists and dieticians in addition to the surgical team. Excellent nursing care is vital. Patients should be adequately optimised before their surgery with attention being paid to their nutritional status and meticulous infection control measures should be in place as many SLE patients have their immunity suppressed by the disease and anti-lupus medications. The presence of antiphospholipid antibodies (prevalence of 30-60% in SLE patients) could result in a coagulopathy that results in a prothrombotic state (despite the coagulation profile suggesting otherwise) or bleeding when associated with very low levels of platelets. Deranged coagulation profiles due to anticardiolipin antibodies are usually not corrected with FFP transfusion⁷. Our patient had an INR of 1.8 which was, however, rapidly corrected with FFP.

The surgical option will depend on the size, location and expertise of the surgeons as in other pancreatic pseudocysts. The main interventions are percutaneous drainage, endoscopic drainage and surgical drainage⁸. While there has been a report of a similar case being managed successfully by percutaneous drainage of the cyst⁹ (probably due to the relatively small size of the cyst), the higher risk for infection associated with this procedure and the size of the cyst worked against such a procedure in this case. Endoscopic drainage could not be performed as the cyst was too low in the abdomen. A Roux-en-Y cystojejunostomy was the preferred definitive drainage procedure in this case because of the cyst size and position as it allows the anastomosis of the Roux loop to the dependent portion of the cyst to allow for maximal drainage (Fig. 1.2).

Figure 2

Figure 1.2



The role of steroids is still unclear in the management of these patients but probably maintenance of the steroid dose or an increase in the dose may be beneficial.

CONCLUSION AND LEARNING POINTS

High index of suspicion of pancreatitis as a possibility is needed in SLE patients complaining of abdominal pain as pancreatitis and pseudocysts can easily be masked by known SLE features and delay in diagnosing pancreatitis can have grave consequences.

The surgical management of these patients poses a challenge as they are immunocompromised, undernourished and coagulation is affected and these problems are further aggravated by steroids and immunosuppressant medications.

Patients should be adequately optimised for better outcome.

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