CASE REPORT

A rare case report of lupus-associated pancreatitis with Evan’s syndrome complicated by huge pseudocyst of pancreas

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Introduction
Systemic lupus erythematosus (SLE) is a multisystem disorder and can affect single or multiple organ systems often resulting in a delayed diagnosis or missed diagnosis. Rarely, acute pancreatitis is the presenting symptom in SLE.[1] The attacks of pancreatitis do not correlate with generalized flare of SLE.[2] Some associate it with the use of corticosteroids[3] and azathioprine.[4,5] Some associate it with vasculitis and thrombosis seen in SLE, but most recently its said to be due to SLE than steroids.[6,7]

Case Report
A 20-year-old female patient known case of SLE with Evan’s syndrome had multiple episodes of acute pancreatitis in 2 years and presented to us with a mass per abdomen [Figure 1]. On examination, the patient had a vertically oval intra-abdominal mass in the left hypochondrium, umbilical region extending into the left lumbar, and epigastric region. The patient also had history of multiple seizure episodes in the past 1 year on treatment with tablet levetiracetam. Patient gives history of macular rash on exposure to sunlight. The patient presented with scarring alopecia over scalp, hyperpigmented skin lesions all over the body. Oral ulcers present over the right side of buccal mucosa. On further evaluation, the patient had megaloblastic anemia, with direct and indirect Coombs test positive, ANA profile positive, and anti-dsDNA titer positive. The patient was on Wysolone 5 mg od and levetiracetam for seizure disorder with severe anemia. After adequate pre-operative management and anemia correction, the patient underwent cystogastrostomy. Due to associated comorbidities and post-operative IV phenytoin infusion, the patient developed purple glove syndrome of the left hand with wrist drop and complete sensory loss. The patient also had cephalic and basilic vein thrombosis and compartment syndrome of the left hand.

Discussion
After appropriate pre-operative evaluation and treatment, the patient was planned for cystogastrostomy. The left Kocher (Chevron rooftop modification) incision was placed and cystogastrostomy was done [Figure 4]. The patient improved symptomatically and was allowed orally on post-operative day 3. On post-operative day 3, the patient developed purple glove syndrome.
syndrome of the left hand with wrist drop and complete sensory loss due to phenytoin infusion which resulted in compartment syndrome, the patient also had cephalic and basilic vein thrombosis. The patient underwent multiple fasciotomies with heparin and dextran infusion. The patient developed bone marrow failure with bleeding manifestations due to ongoing heparin infusion. Aggressive resuscitation was done with blood and fresh frozen plasma transfusion and injection dexamethasone. The patient improved symptomatically with full recovery of the hand function with no residual deformities or gangrenous changes. No abdominal complications were encountered.

**Conclusion**

Here is a rare case report of a young female patient with SLE with Evan’s syndrome with megaloblastic anemia, with seizure disorder with a history of multiple attacks of acute pancreatitis presented with huge pancreatic pseudocyst. This rare case was a great medical and surgical challenge. After working up the case and proper pre-operative management, the patient underwent cystogastrostomy. Postoperatively, we faced an inadvertent complication of purple glove syndrome with superficial thrombosis of the left upper limb which resulted in compartment syndrome, wrist drop, and sensory loss. The patient also had bleeding manifestation postoperatively due to IV heparin with bone marrow failure. Aggressive management of this complication was done with full recovery of the left hand and uneventful abdominal surgery. This case report serves as a future reference and learning opportunity.

**Clinical Significance**

Surgical management of such cases is a real challenge as multiple systems were involved and no consensus exists regarding management of pseudocyst in a case of SLE with Evan’s syndrome. Hence, our case report serves as a learning opportunity and future reference for all other cases.

**References**

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