Case Report

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Recurrent acute pancreatitis in an adult female with sickle cell disease: A case report

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Introduction and Importance: Although risk factors such as cholelithiasis and vaso-occlusive crises exist in sickle cell disease, acute pancreatitis and its recurrence are considered rare complications manifesting as acute abdomen.

Case Presentation: A 33-year-old female with sickle cell disease and established cholelithiasis presented to the center with acute abdomen. After examination, investigation, and contrast enhanced computed tomography, acute pancreatitis was diagnosed. Conservative management was done and cholecystectomy was planned but delayed due to low hemoglobin. In the interval she presented again with similar features and diagnosed with of recurrence. After conservative management and after optimization of patient's hemodynamic status, laparoscopic cholecystectomy was finally performed. Postoperative period and follow-up visit after 2 months were uneventful.

Clinical Discussion: As the features of pancreatitis and vaso-occlusive crisis are similar, the former should be considered as a differential diagnosis of acute abdomen in sickle cell patients. Laparoscopic cholecystectomy is the treatment of choice for symptomatic cholelithiasis in these patients. However, some issues related to management such as preoperative transfusion and prophylactic cholecystectomy are still debated. This uncertainty caused delay in surgery which may have contributed to the recurrence of pancreatitis in our patient. As the risk of recurrence is possible after the first attack, standard guidelines are required for the definite management of the cause.

Conclusion: This case report adds to the limited literature on recurrent acute pancreatitis in sickle cell patients and points out the need for studies on developing management guidelines in such patients and need for prophylactic treatment.

Keywords: case report, laparoscopic cholecystectomy, recurrent acute pancreatitis, sickle cell anemia, sickle cell disease

Introduction

Recurrent acute pancreatitis (RAP) refers to a clinical entity characterized by greater than or equal to two attacks of acute pancreatitis in an individual without any evidence of underlying chronic pancreatitis[1]. Patients with sickle cell disease (SCD) are at risk of developing acute pancreatitis due to ischemic consequence of sickling, or gallstones secondary to chronic hemolysis[2]. Studies have reported the prevalence of cholelithiasis in sickle cell patients, ranging from 11 to 55%[3,4].

However, despite the high risk of developing acute pancreatitis, it is a rare cause of acute abdomen in SCD[5–8] It appears to be more prevalent in the younger population with several cases being reported in this age group[9,10] But only a few cases have been reported in adults[2,11] We found no single report of RAP in the adult population with SCD, however, we came across one case of recurrence in chronic calcific pancreatitis with pseudocyst in a male child[12]

Our case is an interesting one where an adult female, a known case of SCD presented twice to our center with acute abdominal pain and was diagnosed to have RAP. She was managed conservatively for the first two visits and later underwent laparoscopic cholecystectomy.

HIGHLIGHTS

- Recurrent acute pancreatitis is an extremely rare complication of sickle cell disease.
- There is a debate about the status of prophylactic cholecystectomy in sickle cell disease even though cholelithiasis raises the risk of acute cholecystitis, acute pancreatitis in such demographics.
- The definitive management of symptomatic cholelithiasis is complicated by uncertainty about preoperative management of patients.

Method

This case has been reported in line with the Surgical Case Report (SCARE) Criteria[13]

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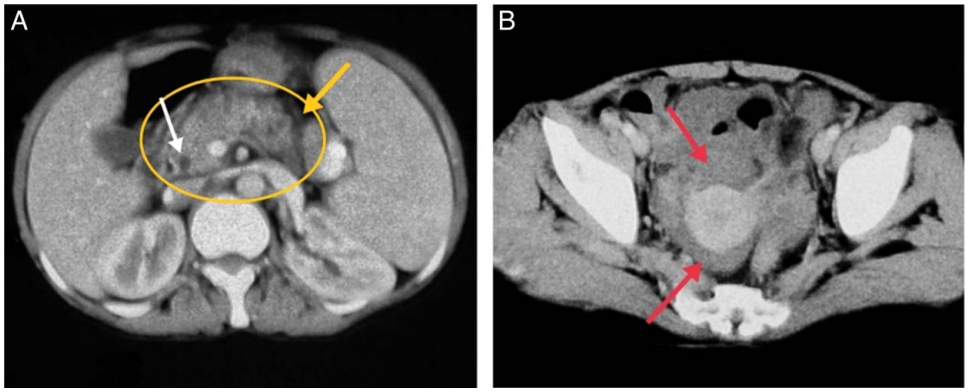

Figure 1. Contrast enhanced computed tomography-abdomen (axial images) showing (A) bulky pancreas (in yellow circle) with peripancreatic fat stranding (marked by yellow arrow), prominent common bile duct (marked by white arrow) and (B) pelvic collection/ascites (marked by red arrows).

Case presentation

Our patient is a 33-year-old married lady from the Tharu community of western Nepal. She is a known case of SCD [homozygous SS, diagnosed via hemoglobin (Hb) High-Performance Liquid Chromatography, with Hb sickle 60.2%]. She presented to our tertiary level hospital, with the complaint of abdominal pain of 2 days duration over the epigastric region with radiation to the back.

She had history of hospital admissions for abdominal pain in the past. Her previous ultrasonography reports revealed the presence of gallstones for which prophylactic laparoscopic cholecystectomy was contemplated on several occasions but was postponed due to episodes of hemolytic crisis. She was under the medications hydroxyurea: 500 mg twice daily, ursodeoxycholic acid: 300 mg twice daily and folic acid: 5 mg once daily, for 3 months and had history of multiple blood transfusions with no documented allergies. Her brother and father are known cases of SCD.

On examination, she was icteric and there was generalized tenderness and guarding of the abdomen. Her spleen was palpable up to the level of the umbilicus. Other systemic examinations were normal.

Following were the values of laboratory investigation during admission: Hb: 10.6 mg/dl, total leukocyte count: 4300/mm^3, differential leukocyte count: $neutrophil_{63,}$ $leukocyte_{30}$, platelets: 7500/mm^3, red blood cells: 2.5 mil/µl, Mean Corpuscular Volume, 99 fl, serum amylase: 1847 U/l, serum lipase: 3373 U/l, serum bilirubin (total/direct): 9.0/2.2, Alkaline Phosphatase: 105 U/l, Alanine Transaminase: 32 U/l, glucose: 131 mg/dl, urea/creatinine: 20/0.5 mg/dl, Na^+/K^+: 142/3.6 mmol/l, serum Lactate Dehydrogenase: 313, reticulocyte count: 5.19%, Coomb's test: negative. The reports showed findings of hemolytic, normocytic anemia with thrombocytopenia and features of obstructive jaundice with raised pancreatic enzymes, suggestive of acute pancreatitis, probably due to obstructive cause.

Contrast enhanced computed tomography of the abdomen was done which demonstrated bulky pancreas with peripancreatic fat stranding and ascites (Fig. 1), yielding a total modified-Computed Tomography Severity Index-score of 6, along with prominent common bile duct (Fig. 1) and cholelithiasis (Figs 2, 3); findings suggestive of acute pancreatitis possibly due to biliary cause.

The differential diagnosis of acute abdomen such as acute appendicitis, acute cholecystitis, and vaso-occlusive crisis (VOC) were considered. A normal leukocyte count and the absence of localizing signs on examination ruled out acute appendicitis and acute cholecystitis. In view of contrast enhanced computed tomography reports a final diagnosis of acute pancreatitis was

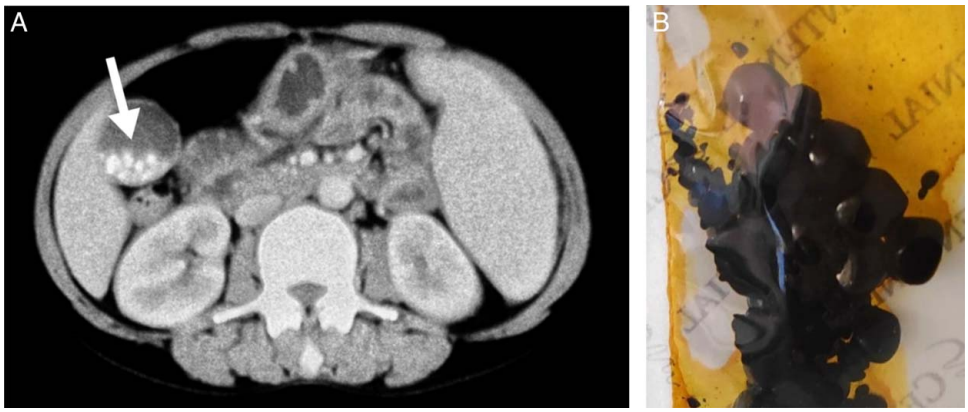

Figure 2. Contrast enhanced computed tomography-abdomen (axial view) showing (A) cholelithiasis (marked by white arrow). To the right (B) is the postoperative specimen showing multiple black pigment stones extracted from the gall bladder.

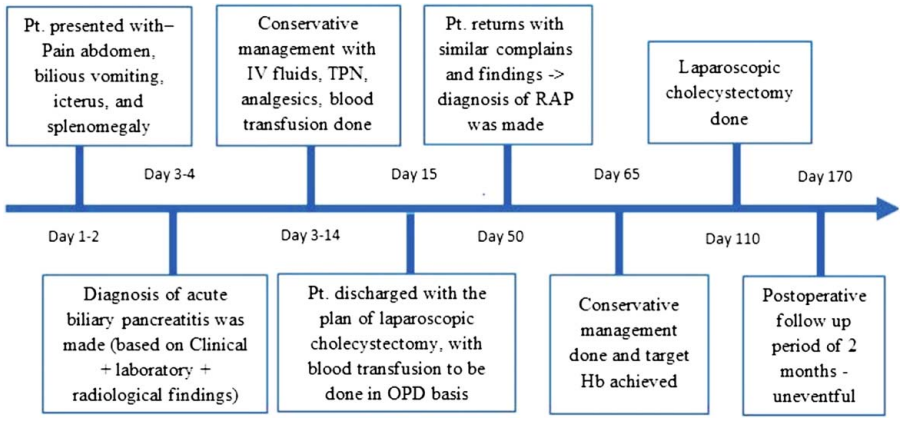

Figure 3. Timeline of events and interventions. Hb, hemoglobin; IV, intravenous; OPD, outpatient department; Pt., patient; RAP, recurrent acute pancreatitis.

made. However, superimposed VOC could not be separately ruled out.

Our patient was initially treated conservatively with intravenous fluids, total parenteral nutrition, analgesics, and blood transfusions. Cholecystectomy was planned but since her Hb levels were constantly low despite blood transfusions, she was discharged with the plan of transfusion on outpatient basis after consulting hematologist and anesthesiologist, with the goal of reaching preoperative target Hb level of 10 g/dl or more prior to surgery.

But within a month of initial treatment, she returned to the hospital with similar symptoms. A diagnosis of RAP was made based on her previous history along with the clinical, laboratory, and imaging findings. Conservative management was done to stabilize the patient. But this time, optimization of Hb level was done with simple transfusion in our center itself and laparoscopic cholecystectomy was performed without delay under general anesthesia by gastrointestinal surgeon with intraoperative finding of a distended gall bladder and common bile duct with a normal Calot's triangle. The cut section of the gall bladder revealed multiple black pigment stones (Fig. 2), the largest one measuring 3 mm in diameter. The postoperative period was uneventful and our patient was symptom free during follow-up at our center 2 months later.

Discussion

An estimated 10% of patients with SCD are hospitalized every year with acute abdomen[2]. Fifty percent of patients in sickle cell crisis present with right upper quadrant pain.

A major concern for the management of acute abdomen in such patients is that the VOC may be indistinguishable from other acute intra-abdominal disease processes, like acute cholecystitis, splenic sequestration, appendicitis, etc.[14]. As such the differential diagnosis of such should be kept in mind.

The initial management of VOC and acute pancreatitis is similar comprising mostly conservative treatment. However, in case of pancreatitis due to underlying causes such as gallstones specific management is required as the risk of recurrence after the first attack is about 20%[11]. This is a major concern as the risk of progression to chronic pancreatitis in patients with at least one recurrence is 35%[15].

The current standard of care for symptomatic gallstones is laparoscopic cholecystectomy, with some reports suggesting that asymptomatic gallstones should be managed similarly in the sickle cell population[16].

Laparoscopic cholecystectomy is considered the gold standard treatment of symptomatic cholelithiasis in patients with SCD because it is safe and results in shorter hospital stays and fewer postoperative complications compared to open operations[17].

Although laparoscopic procedure is widely considered as gold standard treatment in SCD patients, a debate still exists about the preoperative management of patients, especially about the Hb level and preoperative transfusions. Multiple studies have reported target Hb of 10 g/dl or more as the optimum level which reduces postoperative complications, while others claim that it adds risk of transfusion related complications and should be reserved only for necessary cases[18]. This uncertainty regarding preoperative management caused delay in management of the patient which may have led to recurrence in our case to reach optimum Hb level.

Furthermore, the role of prophylactic cholecystectomy in asymptomatic SCD patients has been highly debated with some literature predicting higher costs and increased mortality, while some literature recommends that almost no asymptomatic patients undergo cholecystectomy due to high risk of complication in SCD patients[19,20]. However, in our case prophylactic cholecystectomy could possibly have averted the morbidity.

Our patient was glad that she finally had her gallstones removed, but was concerned about the recurrence of abdominal pain in the future.

The take-away lessons that we can take from this case are that the treatment of asymptomatic cholelithiasis in sickle cell patients might be a better option than dealing with the complications, since laparoscopic cholecystectomy has reduced much of the postoperative morbidity and mortality. Also, in cases where recurrence might be a factor, higher decision-making skills may be required on behalf of the entire team since there is still uncertainty regarding the management of such cases.

Conclusion

Here we report a case where a sickle cell patient with cholelithiasis presented to our center with acute pancreatitis. Initially, conservative management was done and cholecystectomy was planned but since her Hb level was constantly below target, transfusions were done on outpatient basis prior to surgery.

When she presented for the second time with recurrence, we optimized her for surgery, reached target Hb level and performed laparoscopic cholecystectomy without delay. The purpose of this report is to add to the limited literature on RAP and highlight the role of prophylactic cholecystectomy in asymptomatic patients with established cause like cholelithiasis to prevent recurrence. Since the uncertainty in preoperative transfusions caused delay in management during the first visit which may have led to the recurrence in our case, we suggest further research into a revision of management guidelines in such rare cases.

Provenance and peer review

Not commissioned, externally peer-reviewed. This case has not been presented at any conference.

Ethical approval

This is a case report, therefore, it did not require ethical approval from ethics committee.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editor-in-chief of this journal on request.

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The study did not receive any grant from funding agencies in the public, commercial or not-for-profit sectors

Authors' contribution

Aayushi Sharma: conceptualization, writing – original draft. B. K.: writing – review and editing. Anupam Sharma – resources, investigations. K.B.S. – supervision and review. A.N.S. – supervision and review.

Conflict of interest disclosure

The authors report no conflicts of interest.

Research registration

None.

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