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Case Study **Open Access**

Acute Pancreatitis with Normal Amylase/Lipase Levels? A Rare Case Study in Pakistan, an unusual manifestation of cystic fibrosis

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Abstract

Acute Pancreatitis (AP) is the inflammation and auto-digestion of the pancreas and is usually associated with the elevation of serum amylase and lipase levels. Here, we report a rare presentation of AP with normal pancreatic enzymes and lupus vulgaris (LV) in a patient with cystic fibrosis (CF). The chief complaints included severe abdominal pain in association with fever and vomiting. On examination, the abdomen was tense and tender. There were bilateral coarse crepitations with expiratory wheeze more pronounced on the left lower chest. After detailed clinical evaluation, a provisional diagnosis of acute pancreatitis, with pneumonitis and the oral thrush was established. Routine laboratory investigations were performed to confirm the diagnosis. However, the serum amylase and lipase came out within the normal reference range. So, computerized tomography (CT) scan of the abdomen was performed which showed a diffusely swollen and edematous pancreas along with peri-pancreatic stranding, and hence confirmed the diagnosis. Afterward, the patient was managed conservatively, and he had an uneventful recovery. The purpose of reporting this case is to promote awareness among fellow healthcare professionals about this rare manifestation of AP and to prevent any missed diagnosis and serious complications. To conclude, cystic fibrosis may have an unusual presentation like AP without pancreatic enzyme elevation which may create a diagnostic dilemma. Hence, in such cases, a strong clinical suspicion and supportive radiological findings play a critical role in the establishment of the diagnosis.

Keywords: serum amylase and lipase, autosomal recessive, sweat chloride test, mycobacterium tuberculosis, Anti-tuberculous therapy.

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Introduction

Acute Pancreatitis (AP) is becoming an increasingly recognized disorder of all age groups including infants. Hence, AP is considered among the leading causes of admission to hospitals for gastrointestinal tract-related disorders in the United States of America and many other countries[1]. The diagnosis of AP is established if at least two of the following three features are present: distinctive abdominal pain and elevated pancreatic amylase and lipase >3 times the upper limit, and/ or specific imaging findings[2]. Although normal levels of amylase and lipase never exclude AP in a patient presenting with characteristic abdominal pain, this phenomenon is a rarity. The review of medical literature explained that extensive pancreatic necrosis, late presentation, very early stages of pancreatitis, and hypertriglyceridemia might be a cause of normal serum amylase or lipase levels even in the setting of acute pancreatitis[3].

AP is also a frequently encountered clinical entity among the Pakistani population[4,5]. However, we could not find a single Pakistani case report of AP with normal pancreatic enzyme levels. Furthermore, to the best of our knowledge, its concurrent presentation with cystic fibrosis and lupus vulgaris is also an unusual manifestation that has been never reported before. So, we are reporting this case to promote awareness among fellow healthcare professionals about this rare manifestation of AP and to prevent any delayed diagnosis and serious complications.

Case Report

A 9-year-old boy presented with severe dull pain in the epigastric region radiating to the left hypochondrium. It was associated with continuous low-grade fever and vomiting. The child was vaccinated with Bacillus Calmette & Guerin (BCG). However, five years back, he developed pulmonary tuberculosis for which he had completed a six-month course of anti-tuberculous therapy (ATT). But the patient continued to develop recurrent respiratory tract infections. So, a detailed workup was performed, and he was diagnosed with cystic fibrosis, two years ago.

On examination, the patient looked toxic, lethargic, clubbed, and severely malnourished (weighing 16 kg-below 3rd percentile), and had oral thrush. He was febrile (101'F), had tachycardia (134/minute) and tachypnoea (30/minute), and was normotensive (90/60 mmHg). The abdomen was tender with a localized guarding over the left hypochondrium. Murphy's sign was absent along with normal findings on digital rectal examination and gastric lavage. The chest showed bilateral coarse crepitations with expiratory wheeze more pronounced on the left lower chest. Hence based upon history and physical examination, a provisional diagnosis of acute pancreatitis, with pneumonitis and oral thrush, was established. The conservative management was started with antipyretics, intravenous fluids, miconazole oral gel, and oral antibiotics (cefadroxil and sulfamethoxazole-trimethoprim). For bronchospasm, nebulization with a mucolytic agent (N-acetylcysteine) followed by chest physiotherapy was started. The laboratory investigations were performed which are summarized in Table 1.

Table 1: Summary of the various laboratory investigations.

Test	**Normal range**	**Dates**		
CBC		11/06/ 2019	11/08/ 2019	11/11/ 2019
Leukocyte count	4-10 x 10^9 (Neutrophil 43-75%)	**10.6** (Neutrophil 85%)	**10.5** (Neutrophil 72%)	7.2 (Neutrophil 70%)
Haemoglobin	13-18 mg/dl	**10.0**	**10.2**	10.4
Hct	41-45 l/l	32.6	34.3	34.5
MCV	78-96 fl	**74**	**76**	**73**
MCHC	31.5-35.5 g/dl	32.6	30.3	33.1
Platelets	150-400 x 10^3	357	370	320
ESR	Less than 15mm/hour	100	82	46
CRP	Less than 6 mg/dL	96	73	15
Random Blood Sugar	70-140 mg/dL	137	108	
Serum Lactate Dehydrogenase	0.5-1 mmol/L	1.2	0.3	
Serum Electrolytes				
Sodium	135-150 mmol/l	133	137	132
Potassium	3.5-5.0 mmol/l	4.1	5.0	4.2
Calcium	8.5-10.2 mmol/lf	9.1	9.3	9.9

Liver Function Tests				
Serum Bilirubin	0.2-1.2 mg/dl	1.7	0.5	
Alanine Transaminase	40-60 IU/L	10	35	
Aspartate aminotransferase	Less than 60 IU/L	26	12	
Renal Function Tests				
Serum Urea	10-52 mg/dl	14		
Serum Creatinine	0.7-1.2 md/dL	0.3 mg/dL		
Serum Lipase	Up to 40 U/L	**8**	**12**	
Serum Amylase	Up to 90 U/L	**56**	**60**	
Lipid Profile				
S. Cholesterol	Up to 200mg/ dl	193		
Triglycerides	35-165 mg/dl	126		

Unexpectedly, his serum amylase: 56 U/L(12-125) and serum lipase: 8U/L (8-78) were not raised.

Therefore, the computerized tomography (CT) scan of the abdomen was performed which showed a diffusely swollen and oedematous pancreas (>2cm pancreatic band) along with peri-pancreatic fat stranding (typical for AP) as shown in Figure 1 and 2.

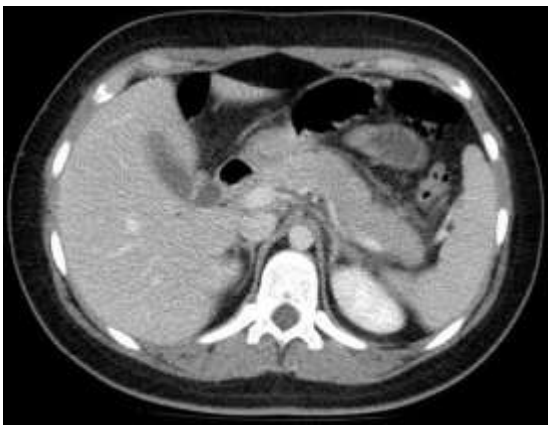

Figure 1: CT abdomen with iodine contrast (portal phase) show diffuse pancreatic enlargement with peripancreatic fat stranding, no pancreatic necrosis or peripancreatic fluid collection.

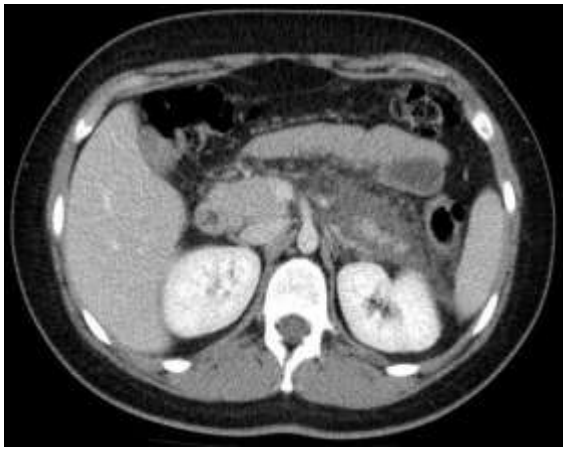

Figure 2: CT abdomen with iodine contrast (portal phase) at level of uncinate process of pancreas clearly show the peripancreatic fat stranding.

The oral antibiotics were changed to intravenous clarithromycin while continuing conservative therapy for AP. The levels of amylase and lipase (60 and 12U/L respectively) remained normal. The follow-up ultrasound after four days showed a pancreatic enlargement with normal-sized pancreaticobiliary ducts. On day five, the enlarged right cervical lymph node developed pustules with scanty pustular discharge. The Tuberculin skin test (TST) was negative as were the gram &Ziehl Nelson stains of pustular discharge. However, due to strong clinical suspicion, fluorescence microscopy was performed which revealed acid-fast bacilli. Hence, the skin lesions were diagnosed as lupus vulgaris. Meanwhile, the patient responded well to medical management and his condition improved. Hence, he was discharged by day seven on his usual treatment of CF and a follow-up plan for lupus vulgaris. The summary of clinical presentation is summarized in Table 2.

Table 2: Summary of clinical presentation.

Date	Summaries from Initial and Follow-up Visits	Diagnostic Testing	Interventions
06-11-2019	**First visit at children Hospital and Institute of Child Health Multan** **Chief Complaints:** The patient presented with acute abdomen along with complaints of fever and vomiting. The patient also had productive cough and dyspnea. **Physical Examination:** A hemodynamically stable patient with bilateral coarse crepitations and expiratory wheeze more pronounced on left side. His abdomen was tender with a localized guarding over the left hypochondrium. He stood below third centile on the weight scale. **Diagnosis:** Acute Pancreatitis and left sided lobar pneumonia. Oral Thrush	Baselines Serum Amylase & Serum Lipase were normal. Plain chest X-ray revealed infiltrates on the left lower zone.	Oral antibiotics, antipyretics and IV fluids. Nebulization with mucolytic agent (N-acetylcysteine) and Chest Physiotherapy. Oral Miconazole gel
11-07-2019	No improvement, rather symptoms worsened over time.	Abdominal CT-scan with IV contrast revealed diffusely swollen pancreas (>2cm pancreatic band) along with peri-pancreatic fat stranding.	Nil per oral IV Fluids, IV clarithromycin,
11-10-2019	Fever, vomiting, cough and bronchospasm was improved but abdominal pain was still present.	The abdominal ultrasound showed a pancreatic enlargement with normal sized pancreatico-biliary ducts.	Same treatment continued.
11-11-2019	The enlarged right cervical lymph node developed pustules with scanty pustular discharge.	The Tuberculin skin test (TST) and gram &Zeil Nelson stains of pustular discharge were negative but fluorescence microscopy revealed Acid Fast Bacilli (AFB). Serum Amylase & Serum Lipase were normal.	Discharged with follow up advice for LV (Gastric Lavage for culture and sensitivity or skin biopsy)
11-20-2019	**First Follow up.** The abdominal pain was significantly improved, and cervical lymph node was having nodular reddish-brown lesions.	Repeated chest X-ray (few infiltrates at the left lower zone).	Anti-tuberculous therapy was started based on the positive skin biopsy
12-20-2020	**Second Follow up.** The patient was asymptomatic with scarring of the cervical node area	Repeated chest X-ray was clear	No side effects of ATT were observed.
01-20-2020	**Third Follow up.** The original wound was barely visible. Monthly follow ups continued until full course of six months		No side effects of ATT were observed.

Discussion

The most common presentation of AP is abdominal pain with elevated serum amylase and lipase usually threefold the normal limit. At the onset of AP, serum amylase increases over 3 to 6 hours with a half-life of 10 to 12 hours and remains elevated for 3 to 5 days. Serum lipase levels increase over 3 to 6 hours peaking in 24 hours and remain elevated for 1 to 2 weeks[6]. Park et al. proved that Lipase has 25% more sensitivity than amylase in the statistical studies where the sensitivity for amylase or lipase (>3 times) combined was only 4% higher than lipase alone[7]. While reviewing the available medical literature we found that only eight cases presented with normal amylase and lipase levels[8]. A growing body of research also showed that isolated increases of these enzymes can be due to a variety of factors, therefore abdominal pain with positive imaging is sufficient to diagnose AP[9]. Hence, normal laboratory tests should be accompanied by an abdominal CT scan with IV contrast (if there is no contra-indication) to establish an accurate diagnosis.

Cystic fibrosis (CF) is inherited as autosomal recessive disease and it has a diverse spectrum of disease as the diagnosis is increasingly being established at adulthood. A typical CF is diagnosed on basis of clinical symptoms and two positive sweat chloride tests on two different occasions[4]. Our patient was labelled as typical CF based on the presence of specific symptoms and two elevated sweat chloride values 4-months apart (96.6 and 93.7 mmol/L).

Cutaneous tuberculosis (CTB) is the tuberculosis of the skin due to the previous sensitization to mycobacterium tuberculosis and a delayed hypersensitivity reaction. It manifests in several phenotypes e.g., lupus vulgaris (LV) is one of its chronic progressive forms. LV is confirmed by laboratory tests (direct demonstration of the acid-fast bacilli either in a tissue smear or biopsy or by isolation in culture or DNA identification by PCR)[5]. Infection with LV results endogenously through the lympho-hematogenous route after reactivation of latent mycobacteria in the immunocompromised state, or rarely via exogenous inoculation[10]. Diagnosis of CTB is at times challenging as the clinical appearance of the lesion may not always be characteristic and may need supportive investigations. In the present case, acid-fast bacilli in the pustular discharge were demonstrated however, TST was negative. This finding could be due to the early disease stage a few numbers of growing TB germs or due to the immunocompromised status of this child.

Conclusion

AP with normal enzymes is a rare possibility that should be investigated further with abdominal CT/MRI scans if the clinical suspicion of AP is high. Further research is needed to better understand the mechanisms underlying the development of AP and the role that these digestive enzymes play.

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