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Case Study

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PROTEIN C AND S DEFICIENCY IN ACUTE MESENTERIC ISCHAEMIA: A CASE REPORT

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ABSTRACT

Protein C and Protein S are vitamin K dependent plasma proteins which are important as anticoagulants. So their deficiency predisposes to conditions associated with increased risk of thrombosis. We present a case of a 50yrs old male presented with abdominal pain in emergency. Provisional diagnosis of acute mesenteric ischaemia was made and patient underwent exploratory laparotomy with subtotal colectomy and ileostomy. Radiological investigations were suggestive of intestinal obstruction Blood investigation showed d-dimer >5000ng/ml, protein C- 62.7% and protein S- 67.7%. Protein S binds and assists activated protein C in the degradation of coagulation factors, therefore their deficiency leads to hypercoagulable state increasing the risk of thromboembolism events. One of these includes acute mesenteric ischaemia; a rare but life threatening condition. Early diagnosis is crucial in such cases for the prompt treatment to improve prognosis and avoid fulminant bowel necrosis.

KEYWORDS: Protein C, Protein S, Thromboembolic events, Acute mesenteric ischaemia.

INTRODUCTION

Protein C and Protein S are vitamin K dependent plasma proteins which are physiological anticoagulants. Their deficiencies are rare and predispose to a hypercoagulable state increasing the risk of thromboembolic events, often at atypical sites. One such consequence being mesenteric ischaemia presenting as acute abdomen with intestinal obstruction, making it difficult to diagnose and manage the underlying coagulopathy.

Case

A 50 years old male presented with abdominal pain in the emergency. Pain was sudden in onset, and diffuse and non-radiating in nature, aggravated after meal and relieved on medication. He had history of non- passage of stool and flatus for two days. Patient had no past history of surgery but had been a smoker for 20years. On examination he was conscious, oriented and afebrile. His pulse rate was 102/min and blood pressure 100/60mmHg. On per abdominal examination, his abdomen was distended and tender and bowel sounds were absent. His blood investigation showed border line hemoglobin and hematocrit with leukocytosis (table1) and altered coagulation profile (table2). Arterial blood gas analysis was indicative of metabolic acidosis (table1). The metabolic profile, serum amylase and lipase values were within their normal reference ranges. Electrocardiogram and echocardiography was normal. Radiological findings were suggestive of intestinal obstruction (figure 1 and 2). Resected specimen on histopathology showed liquefactive necrosis of colon. Other investigations like pus culture and urine culture showed no growth. Provisional diagnosis of acute mesenteric ischaemia(AMI) was made and patient underwent exploratory laparotomy with subtotal colectomy and ileostomy.

Postoperative period was uneventful and patient was discharged after successful anticoagulation and prescribed with Coumadin. The coagulation profile was repeated after 6 weeks and Protein C and S levels were still found low.

Table 1: Blood investigation showing hemogram, biochemical profile and ABG analysis.

SrNo	Investigation	Result
1	Hemoglobin	12.8 gm
2	TLC	13.4×10^9/L
3	DLC	N86/L62/M1/E1
4	Hct	36.4 %
5	pH	7.237
6	pO_2	63.1 mmHg
7	pCO_2	49.5 mmHg
8	HCO_3^-	20.6 mEq/L
9	BE	-6.8

Table 2: Coagulation profile.

SrNo	Investigation	Result
1	PT	12.5 seconds
2	INR	1.08
3	D-Dimer	>5000 ng/ml
4	Protein C	62.7 %
5	Protein S	67.7 %

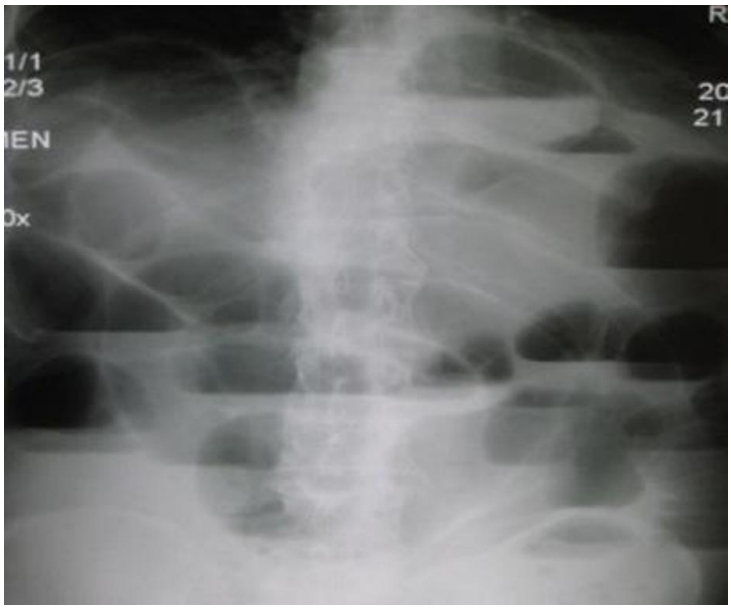

Figure 1: Abdominal X-Ray (erect) showing multiple air-fluid level.

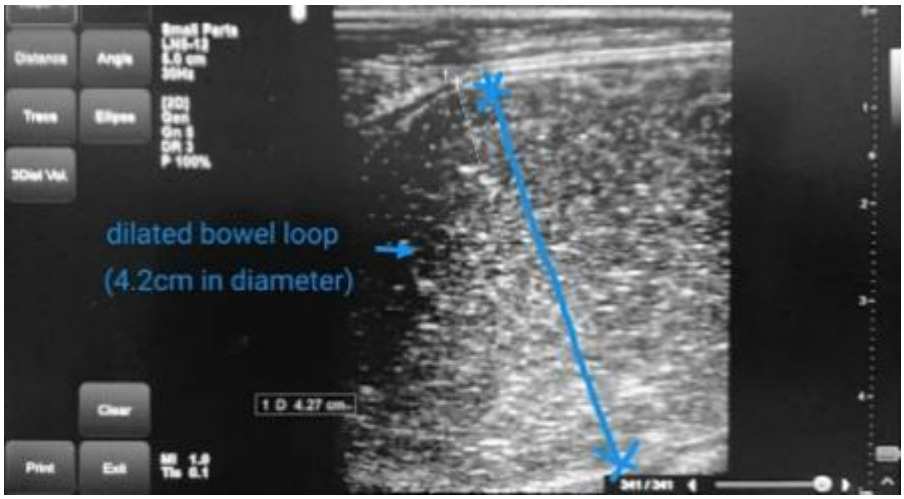

Figure 2: USG abdomen showing dilated bowel loop.

DISCUSSION

AMI, a rare cause of acute abdomen, is a life-threatening vascular emergency which demands early diagnosis and intervention to adequately restore mesenteric blood flow and to prevent bowel necrosis and patient death. The final common pathway of all the specific causes of mesenteric ischaemia is bowel infarction where the patient has peritoneal signs, hemodynamic instability, and signs of sepsis with multi-organ failure.[1]

Here patients present with non-specific symptoms, making the diagnosis difficult and delays the treatment. It can only be suspected in presence of supporting past or family history of thrombo-embolic events, drugs etc. and precipitating causes such as past surgery, smoking, atrial fibrillation should not be neglected. While evaluating the patient workup for genetic factor should also be done. Nearly, 25% of patients may have normal findings on abdominal radiography and hence does not rule out AMI. Contrast-enhanced computed tomography (CECT) has been found very useful in diagnosing mesenteric ischaemia and its various causes like arterial occlusion, venous occlusion, strangulating obstruction and also in differentiating it from other causes of acute abdomen like acute pancreatitis, acute appendicitis etc. However, it still can detect about 90% of cases.[2] Many get diagnosed only during surgery. Thus, laboratory investigations like Protein C and Protein S estimation are important for evaluation of such cases and prompt management.

Clinical evidence of Protein C involvement in the regulation of coagulation comes from the observation that low levels of Protein C are associated with recurrent familial thrombosis.[3] Baburajetal. reported a case of acute abdomen later diagnosed to have Protein S deficiency and managed with anticoagulants.[4] Another case of acute mesenteric thrombus with Protein C deficiency was reported by Dina et al.[2] Recent studies state that Protein C and Protein S deficiency increases the risk the arterial blockages in young individuals. A study of young Indians showed that Protein C deficiency alone or with Protein S deficiency is significantly associated with ischemic stroke in young adults.[5]

Protein C, after getting activated, inhibits coagulation by degrading activated factors V and VIII. Protein S binds and assists activated protein C in the degradation of coagulation factors. Protein C deficiency and Protein S deficiency can be both genetic and acquired. The genes for Protein C - PROC and Protein S - PROS1 are located at chromosome 2q 14.3 and 3q11.1 respectively have autosomal dominant inheritance and prevalence of 1 in 500 population.[6,7] Protein C deficiency is more common of which Type I is more common than type II.[6] Before diagnosing patient with inherited PC or PS deficiency, its deficiency due to acquired causes like liver disease, vitamin K deficiency needs to be ruled out and test should be repeated with interval of 4-6weeks.[2] Studies have shown that smoking leads to acquired activated protein C deficiency (dose-dependent hypercoagulability) which in turn increases the risk of thrombosis (here presenting as AMI). This acquired deficiency due to low circulating levels of activated protein C is more profound in male smokers.[8] A similar case of a male smoker was reported who was radiologically diagnosed with AMI and given thrombolytic therapy.[9]

To conclude, patients presenting to emergency with acute abdomen may have a range of underlying causes, of which acute mesenteric ischaemia is rare and can be easily missed. The estimation of these coagulation profile parameters will aid in the early recognition and prompt management of the condition therefore preservingthe bowel viability and thus lessen the complications.

REFERENCES

1. Oldenburg WA, Lau LL, Rodenberg TJ, Edmonds HJ, Burger CD. Acute mesenteric ischaemia: a clinical review. Archives of internal medicine, 2004; 164(10): 1054-62.
2. Rharrit D, Harmouche H, Baroudi S, Mezalek ZT, Adnaoui M, Aouni M, et al. Protein C deficiency and mesenteric venous thrombosis. Canadian Journal of Surgery, 2009; 52(2): E35.
3. Honghong Li (2015) Two Cases of Recurrent Vascular Events Due to Protein C Deficiency. J ClinToxicol, 5: 243.
4. Baburaj K, Salma Hanam A. Incidence of mesenteric arterial thrombosis in protein S deficiency patients. IAIM, 2015; 2(11): 74-77.
5. Patel ML, Sachan R, Seth G. Combined deficiency of proteins C and S: ischaemic stroke in young individuals. Case Reports, 2013; 2013: bcr2012008016.
6. Bharadiya AA, Aundhakar SC, Panpalia NG, Jaju JB. Protein C and S deficiency presenting as acute abdomen. Medical Journal of Dr. DY Patil University, 2015; 8(1): 48.
7. Lipe B, Ornstein DL. Deficiencies of natural anticoagulants, protein C, protein S, and antithrombin. Circulation, 2011; 124(14): e365-8.
8. Fernández JA, Gruber A, Heeb MJ, Griffin JH. Protein C pathway impairment in nonsymptomatic cigarette smokers. Blood Cells, Molecules, and Diseases, 2002; 29(1): 73-82.
9. Sharma M, Kumawat G. Celiac artery thrombosis and superior mesenteric artery stenosis. Heart India, 2016; 4(4): 136.