

## CASE REPORT

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# Haemolytic Uraemic Syndrome Following Acute Pancreatitis

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### ABSTRACT

**Context** Haemolytic uraemic syndrome is a common cause of renal failure in children but it is a rare condition in adults. Acute pancreatitis in adult as a cause of haemolytic uraemic syndrome is very rare.

**Case report** A 19-year-old male presented with symptom and signs suggestive of acute pancreatitis which was confirmed as his serum amylase was significantly raised. Within three days of admission he developed acute renal failure with evidence of haemolytic anaemia and thrombocytopenia. A clinical diagnosis of haemolytic uraemic syndrome was made and he was treated with plasma exchange. He made a complete recovery.

**Conclusion** Renal failure in a patient with acute pancreatitis is rarely due to haemolytic uraemic syndrome. But it is important to consider this differential diagnosis so that early treatment can be instituted to prevent mortality.

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### INTRODUCTION

Haemolytic uraemic syndrome (HUS) is characterised by microangiopathic anaemia with thrombocytopenia and acute renal failure. HUS precipitated by E. Coli O157:H7 induced diarrhoea is one of the most common cause of renal failure in infants and children. But HUS is a rare disease in adult. Acute

pancreatitis as a result of HUS is rare and affects about 2% of adults suffering from HUS but HUS as a result of acute pancreatitis has only been discussed in a few case reports [1].

We describe a further case of acute pancreatitis highlighting the importance of early diagnosis and appropriate management.

### CASE REPORT

A 19-year-old male was admitted to the Surgical Unit through Emergency Unit. He presented with one-day history of severe upper abdominal pain, associated with frequent vomiting. There was no history of diarrhoea or fever. There was no past history of any significant illness. On examination he was found having epigastric tenderness. His vital parameters were normal and blood test at the time of admission revealed normal haemoglobin, full blood count, urea, electrolytes, and liver function tests. Serum amylase was raised to 1,426 IU/L (reference range: 25-125 IU/L) and a diagnosis of acute pancreatitis was made. Ultrasound revealed oedematous body and tail of pancreas, biliary tree was not dilated and there was sludge in the gallbladder.

In next two days his renal function progressively deteriorated and on 3<sup>rd</sup> day of his admission his urea and creatinine were 21.1 mmol/L (reference range: 0-7.5 mmol/L) and 590 µmol/L (reference range: 0-120 µmol/L) with associated oligurea. He was shifted to ICU. First of all cause of his renal failure was thought to be pancreatitis

associated hypovolemia but it was noticed that his haemoglobin level had dropped to 8.8 g/dL from 14.9 g/dL at the time of admission. His clotting profile was normal and there was evidence of thrombocytopenia with platelet count of  $32 \times 10^9/L$  (reference range:  $15-40 \times 10^9/L$ ). His clotting profile was normal and a normal fibrin degradation product (FDP) rule out disseminated intravascular coagulation. In view of his renal failure he was seen by the nephrologists and a provisional diagnosis of HUS was made. Further investigation revealed evidence of intravascular haemolysis with raised reticulocyte count of  $60,000 \text{ mm}^{-3}$  (reference range:  $25,000-120,000 \text{ mm}^{-3}$ ), schistocytes in blood smear and haptoglobin at 0.1 g/L (reference range: 0.2-2.4 g/L). The Coombs' test was negative. A renal biopsy revealed thrombi in few capillary loops, tubules showed acute tubular damage with inflammatory infiltrate and interstitial oedema. Amyloid was not seen and immunohistochemistry for IgA, IgG and IgM was negative.

He required regular haemodialysis and fresh frozen plasma transfusion. His renal function progressively improved and he was discharged from ICU to ordinary ward on day 30 and was discharged from hospital after 2 months of admission.

## DISCUSSION

Acute renal failure in patients with acute pancreatitis carries a poor prognosis and its incidence varies with the severity of pancreatitis with incidence up to 42% has been reported [2]. In a retrospective study of 563 patients with acute pancreatitis the incidence of renal failure was 14% [3]. The renal failure in our case of acute pancreatitis was part of HUS.

Aetiology of HUS in adult is more heterogeneous compared to HUS in children where 80% of the cases are associated with E. Coli O157:H7 infection [4, 5]. The overall prognosis of HUS in adult is poor compared to children [6] and rate of chronic renal

failure after HUS ranged from 40 to 60% in adult series [4, 7]. Thus it is important to consider the diagnosis of HUS in patients with renal failure in case of pancreatitis.

The mechanism of development of HUS following pancreatitis is not clear. As there are only 20 cases reported in the literature [1], there has not been enough opportunity to understand the aetiology. Several hypotheses has been put forward including the role of TNF-alpha and IL-1 which are important mediators for pancreatitis and might induce widespread vascular endothelial injury [8]. The other suggestion is modification of circulating Von Willebrand molecules by pancreatic proteases, which leads to platelet aggregation [9]. Acquired or congenital deficiency of Von Willebrand factor has been shown to be the underlying cause of thrombotic angiopathies. Recently serum measurement of von Willebrand factor cleaving protease (ADAMTS-13) has been used to differentiate between thrombotic thrombocytopenic purpura (TTP) and HUS. As patients with TTP have little or no ADAMTS-13 activity in plasma compared to patients with HUS [10, 11]. It has also been suggested that patients with ADAMTS-13 activity might have better prognosis than patients without ADAMTS-13 [12].

The histological heterogeneity of haemolytic uraemic syndrome has been already well demonstrated [13, 14]. The typical lesion in TTP shows widespread vascular thrombosis in glomerular capillaries but in HUS the renal lesion may only show glomerular, tubular or vascular lesion [13]. As in our case there was evidence of tubular inflammation without wide spread thrombosis in vessels.

The treatment of HUS following acute pancreatitis involves supportive measures including haemodialysis if required as in our case. It has been suggested that fresh frozen plasma infusion improves the renal outcome in adult HUS [15]. Several retrospective studies in adults suggested that plasma therapy may improve mortality and renal recovery in patients with HUS [4, 16].

## CONCLUSION

It is important that clinicians treating acute pancreatitis should be aware of HUS as a possible complication of acute pancreatitis and in any patient with acute pancreatitis developing renal failure HUS must be ruled out by blood tests. Early institution of plasma therapy will improve prognosis and long term renal function.

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**Keywords** Hemolytic-Uremic Syndrome; Kidney Failure; Pancreatitis, Acute Necrotizing

**Abbreviations** FBC: full blood count; fibrin degradation product; HUS: haemolytic uraemic syndrome; TTP: thrombotic thrombocytopenic purpura

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## References

1. Boyer A, Chadda K, Salah A, Bonmarchand G. Thrombotic microangiopathy: an a typical cause of acute renal failure in patients with acute pancreatitis. *Intensive Care Med* 2004; 30:1235-9. [PMID 15069598]
2. Herrera Gutierrez ME, Sellar Perez G, de La Rubia De Gracia C, Chaparro Sanchez MJ, Nacle Lopez B. Acute renal failure profile and prognostic value in severe acute pancreatitis. *Med Clin (Barc)* 2000; 115:721-5. [PMID 11141437]
3. Kes P, Vucicevic Z, Ratkovic-Gusic I, Fotivec A. Acute renal failure complicating severe acute pancreatitis. *Ren Fail* 1996; 18:621-8. [PMID 8875688]

4. Hollenbeck M, Kutkuhn B, Aul C, Leschke M, Willers R, Grabensee B. Haemolytic-uraemic syndrome and thrombotic-thrombocytopenic purpura in adults: clinical findings and prognostic factors for death and end-stage renal disease. *Nephrol Dial Transplant* 1998; 13:76-81. [PMID 9481719]
5. Matsumae T, Takebayashi S, Naito S. The clinico-pathological characteristics and outcome in hemolytic-uremic syndrome of adults. *Clin Nephrol* 1996; 45:153-62. [PMID 8706355]
6. Remuzzi G. Hemolytic uremic syndrome: past and present. *Am J Kidney Dis* 2000; 36:LIV-VI. [PMID 11054374]
7. French Cooperative Study Group for Adult HUS. Adult hemolytic uremic syndrome with renal microangiopathy. Outcome according to therapeutic protocol in 53 cases. *Ann Med Interne (Paris)* 1992; 143(Suppl 1):27-32. [PMID 1300883]
8. Silva VA. Thrombotic thrombocytopenic purpura/hemolytic uremic syndrome secondary to pancreatitis. *Am J Hematol* 1995; 50:53-6. [PMID 7668225]
9. Daryanani S, Wilde JT. Relapsing thrombotic thrombocytopenic purpura in association with recurrent pancreatitis. *Clin Lab Haematol* 1998; 20:317-8. [PMID 9807681]
10. Furlan M, Lammle B. Deficiency of von Willebrand factor-cleaving protease in familial and acquired thrombotic thrombocytopenic purpura. *Baillieres Clin Haematol* 1998; 11:509-14. [PMID 10097823]
11. Furlan M, Robles R, Galbusera M, Remuzzi G, Kyrle PA, Brenner B, et al. von Willebrand factor-cleaving protease in thrombotic thrombocytopenic purpura and the hemolytic-uremic syndrome. *N Engl J Med* 1998; 339:1578-84. [PMID 9828245]
12. Haspel RL, Jarolim P. The 'cutting' edge: von Willebrand factor-cleaving protease activity in thrombotic microangiopathies. *Transfus Apheresis Sci* 2005; 32:177-84. [PMID 15784452]
13. Pourrat O, Touchard G, Robert R, Badia P, Bauwens M, Hauet T, Patte D. A kidney biopsy is clearly mandatory to confirm the indication of plasma exchanges in adult haemolytic uraemic syndrome. *Ann Med Interne (Paris)* 1994; 145:369-72. [PMID 7985952]
14. Taylor CM, Chua C, Howie AJ, Risdon RA. Clinico-pathological findings in diarrhoea-negative haemolytic uraemic syndrome. *Pediatr Nephrol* 2004; 19:419-25. [PMID 14986082]
15. Tostivint I, Mougnot B, Flahault A, Vigneau C, Costa MA, Haymann JP, Sraer JD, et al. Adult haemolytic and uraemic syndrome: causes and

prognostic factors in the last decade. *Nephrol Dial Transplant* 2002; 17:1228-34. [PMID 12105245]

16. Lara PN Jr, Coe TL, Zhou H, Fernando L, Holland PV, Wun T. Improved survival with plasma exchange

in patients with thrombotic thrombocytopenic purpura-hemolytic uremic syndrome. *Am J Med* 1999; 107:573-9. [PMID 10625026]

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