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ANSWER TO THE PHOTO QUIZ

Fatal haemolytic shock

Keywords - clostridium perfringens, haemolysis, septic shock

Diagnosis: *Clostridium perfringens* liver abscess

Computed tomography of the abdomen shows multiple pyogenic liver abscesses. Most pyogenic liver abscesses are caused by enteric gram-negative bacilli of which *Escherichia coli* and *Klebsiella pneumoniae* are cultured most frequently. In only 1.2% of all patients with liver abscesses is *Clostridium perfringens* the causative organism.[1]

C. perfringens is an anaerobic, gram-positive, rapid-growing bacillus which produces extracellular enzymes and toxins. *C. perfringens* can be found in soil and food, and can be a pathogen as well as a member of the normal human gastrointestinal tract.[2] Infections mostly originate from the uterus, colon or biliary tract and may be fatal. *C. perfringens* has an enormously short generation time of 8-12 minutes. Therefore, although rarely seen, acute infections can progress quickly and can result in the swift development of necrotising soft tissue infection, liver abscesses and septic shock.[3] This septic shock is partially mediated by cytokines as well as the production of toxins that cause diffuse cellular necrosis. In addition, patients may develop massive intravascular haemolysis, which occurs in 7% to 15% of cases. Haemolysis is mediated by the alpha toxin lecithinase, which has phospholipase and sphingomyelinase activities. This toxin induces hydrolysis of phospholipids, which causes disruption of the red blood cell membrane. Haemolysis is associated with a high mortality of up to 80%, occurring within a median time of eight hours from admission to death.[3,4] Furthermore, the alpha toxin averts pathogen clearance by modulating the immune response and causes a microaerophilic environment by vasoconstriction as well as induction of local coagulation and haemostasis leading to rapid proliferation of *C. perfringens*. Early administration of antibiotics and surgical intervention are paramount and improve survival.

Our patient presented with severe haemolysis; the haemoglobin levels dropped within two hours from 6.8 mmol/l to 0.9 mmol/l and the patient needed massive blood transfusion. Other laboratory tests, such as haptoglobin, lactate dehydrogenase and bilirubin, could not be determined because of the degree of haemolysis (spherocytes 5-20%, H-index 2964). The patient rapidly developed septic shock, without any signs of improvement after percutaneous drainage, empiric broad-spectrum antibiotics (meropenem, penicillins and vancomycin) and vasopressor therapy. Gangrenous cholecystitis was found during a laparotomy and was considered to be the source of infection. Twenty-four hours after the patient died, blood cultures were found to be positive for *C. perfringens*. This fatal case illustrates the fulminant course and emphasises the importance of early recognition during infection with *C. perfringens*, as well as the negative prognostic marker of haemolysis in this infection.

Disclosure

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References

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