

Case Study

Cryptogenic Invasive Klebsiella Pneumoniae Liver Abscess Syndrome (CIKPLA) – Case Report and Review of Literature

Abstract:

Klebsiella pneumonia is a common bacterial pathogen that is known to cause diverse community acquired and nosocomial infections^{1, 2}. These infections commonly occur in hospitalized individuals with impaired host defenses^{3}, however the proportion of community acquired vs nosocomial infection has markedly increased in recent years^{4}. A community acquired syndrome of cryptogenic invasive Klebsiella Pneumoniae Liver Abscess Syndrome (CIKPLA) has been known to typically affect individuals of Southeast Asian origin, and is complicated by devastating septic spread to other organs^{5}. We report a case of community acquired invasive Klebsiella infection with multi organ involvement in an individual of non-Asian origin suggesting a need for increased vigilance for early diagnosis and prompt management to prevent disastrous sequelae.

Case Report:

A 64 year old Sudanese male with a background of hypertension and diabetes mellitus for 13 years presented with history of low grade fever and productive cough of 10 days duration. 2 days after the fever, the patient developed pain in the right ankle along with redness and mild pain and swelling in the left calf. He visited a private clinic and received amoxicillin for 5 days without resolution of his symptoms. On the day of admission he developed dull aching, continuous, non- radiating pain on the right side of the abdomen, unrelated to food intake and without alteration in bowel or bladder habits, nausea, vomiting or jaundice along with urinary discomfort. The following day he also developed pain, redness and photophobia along with sticky, mucoid discharge from his right eye.

On physical examination, he had a regular pulse of 80 beats/min, a respiratory rate of 17 breaths/min, temperature of 38.2 °C and a blood pressure of 145/90mmHg. The other notable physical findings were distention of neck veins. Pitting pedal edema was present bilaterally up to the mid-calf region. There was a red, warm, tender swelling over the right medial malleolus measuring 7 X 5 cm with minimal purulent discharge. Abdominal examination revealed tenderness in the right upper quadrant and a palpable liver edge of 4cm. Respiratory examination revealed decreased breath sounds and crackles over right basal area. The right eye was chemosed with conjunctival and ciliary injection along with mucoid discharge. The left eye appeared normal. Visual acuity was CF at 1 meter & at 5 meters for right and left eye, respectively. Pupil was sluggishly reactive on the right side and normal on the right. Extra ocular movements were normal bilaterally. Other systems were unremarkable.

His initial laboratory tests showed a platelet count of 132×10^3 U/L with a normal white cell count and hemoglobin. The serum sodium was 125mmol/L, bicarbonate of 18mmol/L and blood glucose of 23.42mmol/L with rest of the electrolytes in reference range. Of note was a serum albumin of 26g/L, ALT of 92U/L, ALP of 225 with a normal AST, lipase and bilirubin. Blood cultures were positive for *Klebsiella pneumoniae* resistant only to ampicillin. Culture from the urine sample and the leg ulcer also grew the same organism. Ultrasound (Figure 2) abdomen revealed a liver abscess. A CT guided percutaneous drain was yielding 250cc of purulent fluid, with *Klebsiella pneumoniae* isolated from cultures of the aspirated pus. The patient was initially started on Piperacillin/Tazobactam and urgent ophthalmology consultation was sought. Despite the initial antibiotic treatment, the patient had worsening eye symptoms with tense proptosis and conjunctival chemosis. The patient was diagnosed with endogenous ophthalmitis and total retinal detachment. CT scan showed orbital cellulitis. The antibiotic was changed to Meropenem and IV steroids were added. Unfortunately he developed globe perforation of right eye and underwent evisceration under general anesthesia. The patient's general condition improved after this. He completed the course of antibiotics and was subsequently discharged.

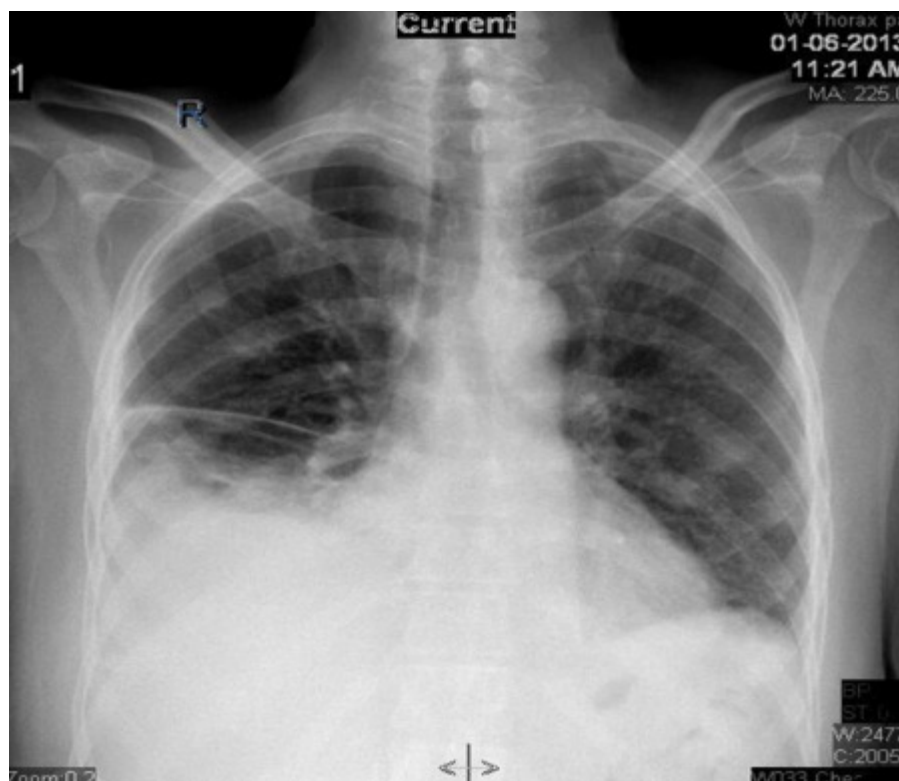


Figure 1, Chest X-ray showing a heterogeneous opacity involving the mid and lower zone on the right side along with obliteration of the right Cardiophrenic angle, suggestive of pleural effusion



Figure 2, Ultrasound scan of abdomen showing hepatomegaly (17cm) with multiple echogenic foci likely representing aerobilia probably as a result of gas forming microorganism with abscess, without any obvious fluid.

DISCUSSION

Klebsiella pneumoniae is an encapsulated Gram-negative bacillus which belongs to the family of Enterobacteriaceae. It is known to cause diverse community-acquired and nosocomial infections {1} and presents with distinct clinical and epidemiological features throughout the world. It is also known to be associated with gastrointestinal infections {6}

Klebsiella pneumoniae has emerged as a major cause of primary or cryptogenic liver abscess, along with an increasingly recognized condition known as Cryptogenic Invasive *Klebsiella Pneumoniae* Liver Abscess Syndrome (CIKPLA). It is commonly found in Diabetics – possibly due to their impaired phagocytosis mechanism of encapsulated organisms {7, 8}. Other than diabetes, invasive *Klebsiella* infections have been also found in individuals with impaired host defenses (such as chronic alcoholism, malignancy, hepatobiliary disease, chronic obstructive pulmonary disease, glucocorticoid therapy, and renal failure) {4, 9-14}, suggesting intrinsic virulence factors enabling the organism to escape host immunity {4}. K1 and K2 capsular strains, coded by the *rmpA* genes have been attributed to the invasive and virulent potential of *Klebsiella pneumoniae* {15}, with K1 strains being significantly associated with pyogenic liver abscess and endogenous endophthalmitis {16,17}. However, to date, its exact pathological mechanisms remain unclear {2}. Although the *Klebsiella pneumoniae* organism was not serotyped for the presence of *magA* gene and K1 capsular type in our patient, he demonstrated poor prognostic features suggesting a virulent form of strain.

CIKPLA Syndrome is frequently associated with one or more complications such as meningitis, endophthalmitis, lung abscess, or fasciitis and has been commonly reported in Taiwanese population

{18}, with a few reports emerging in United States (of Asian descent) {19}. This syndrome has never been reported in Middle Eastern or Sudanese population.

A few studies have analyzed poor prognostic features associated with CIKPLA syndrome. These include a Glasgow coma scale < 7 - prior to initiation of treatment {20}, thrombocytopenia, raised white cell counts and reduced CSF glucose levels {21}. Treatment of choice for CIKPLA syndrome includes third generation Cephalosporin, preferably Ceftriaxone

Endogenous endophthalmitis, caused by *K. pneumoniae* liver abscess is often a devastating septic metastatic infection. It has mostly been described in association with primary liver abscess in patients from East Asia, particularly Taiwan {8, 22-26, 32} and to a much lesser extent in other parts of the world. After diagnosis pyogenic liver abscess, occurrence of ocular symptoms within 48-72 hours suggests the possibility of septic endophthalmitis via hematogenous spread from the liver abscess {23}. Vigorous search for an intraocular infection should be considered in patients presenting with *Klebsiella pneumoniae* liver abscess. Considered to be rare previously, *Klebsiella pneumoniae* endophthalmitis accounts for 2 – 8% of all endophthalmitis cases {16}

We highlight a case of Sudanese male with CIKPLA syndrome demonstrating liver abscess, pneumonia, urosepsis, cellulitis and endophthalmitis with total retinal detachment. The risk factors in our patient included poorly controlled diabetes and chronic heavy alcohol consumption. Despite intensive therapy by an ophthalmologist, visual outcome was poor in our patient. The aim of this case report is to alert clinicians about increasing prevalence of CIKPLA syndrome, its different clinical presentations and occurrence in different populations. We suggest a need for increased vigilance while managing such cases and regularly performing eye examinations at the time of diagnosis so as prevent serious debilitating disease complications.

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