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SCIENTIFIC LETTER

Invasive liver abscess syndrome with septic pulmonary embolisms☆

Síndrome de absceso hepático invasivo con embolismos pulmonares sépticos

Klebsiella pneumoniae (*K. pneumoniae*) is a Gram-negative bacillus that causes nosocomial pneumonia and intra-abdominal and urinary tract infections[1]. Liver abscesses in patients with *K. pneumoniae* infection were first described in Taiwan in 1980. *K. pneumoniae* liver abscesses generally present with extrahepatic complications, such as bacteremia, sepsis, metastasis to the central nervous system (CNS), endophthalmitis, and necrotizing fasciitis, comprising the clinical syndrome now defined as invasive liver abscess syndrome[2]. The metastatic infection rate varies from 3.5 to 20%, and the mortality rate from 2.8 to 10.8%[3]. The hypervirulent serotypes, K1 and K2, have been identified and are characterized by increased capsule production and siderophore expression that worsen the clinical presentation. Hypervirulent *K. pneumoniae* has a mucoid phenotype and the associated genes include the regulator of mucoid phenotype A (rmpA), rmpA2, and the mucoviscosity-associated gene A (magA) that increases capsule production[4].

A 23-year-old woman, with a history of type 2 diabetes mellitus, arrived at the emergency room with diffuse abdominal pain, nausea, vomiting, and fever of 3-day progression. The pain intensified, radiating to the epigastrium and right hypochondrium. Her vital signs were heart rate of 131 beats per minute, respiratory rate of 24 breaths per minute, blood pressure of 110/60 mmHg, and temperature of 38.6 °C. Physical examination revealed bibasilar crackles, diffuse abdominal pain upon palpation, and signs of peritoneal irritation. Laboratory test results reported leukocytosis of 19,280 cell/mm^3, neutrophils of 79.8%, and admission glucose level of 489 mg/dl. The rest of the laboratory tests were normal. A chest x-ray was ordered and identified multiple bilateral infiltrates (Fig. 1). A non-contrast chest and abdominal computed tomography (CT) scan showed a large 6 × 7.1 × 3 cm ruptured liver abscess,

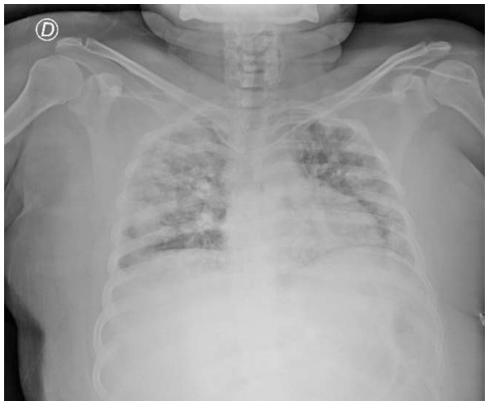

Figure 1 Chest x-ray with multiple bilateral infiltrates.

with gas bubble production, located in the right liver lobe. There was no diaphragmatic rupture or signs of continuity to the chest. Pneumoperitoneum, with free fluid in both parietocolic gutters, was present (Fig. 2A). Multiple cavitated pulmonary nodules, consistent with septic embolisms, were reported in the chest (Fig. 2B). The patient had not been previously treated, and so broad-spectrum antibiotics were started, but her clinical condition rapidly worsened and she progressed to septic shock, requiring vasopressors. The patient was taken to the operating room, where emergency laparotomy was performed, revealing approximately 1 liter of purulent matter in the abdominal cavity. Pus samples were taken for culture. Liver biopsy was performed to rule out malignancy. The abscess identified in segment IV was drained and the critically ill patient was admitted to the intensive care unit. She died 10 h after the surgical procedure. The culture report stated *K. pneumoniae* and the liver biopsy described fibrinous and purulent material.

Over the past few decades, the association of *K. pneumoniae* with a complex invasive syndrome has become recognized and is now identified as invasive liver abscess syndrome. It is a disease with worldwide prevalence and the frequency of *K. pneumoniae* in pyogenic liver abscesses has increased over the past 30 years in Taiwan[2]. The disease presents in immunocompromised patients, and diabetes mellitus is the most common predisposing factor[5]. A lower frequency of metastasis has been observed in diabetic patients with strict glycemic control[6]. Metastatic disease is associated with severe clinical presentation and a high mortality rate. Chang et al.[3] reported that

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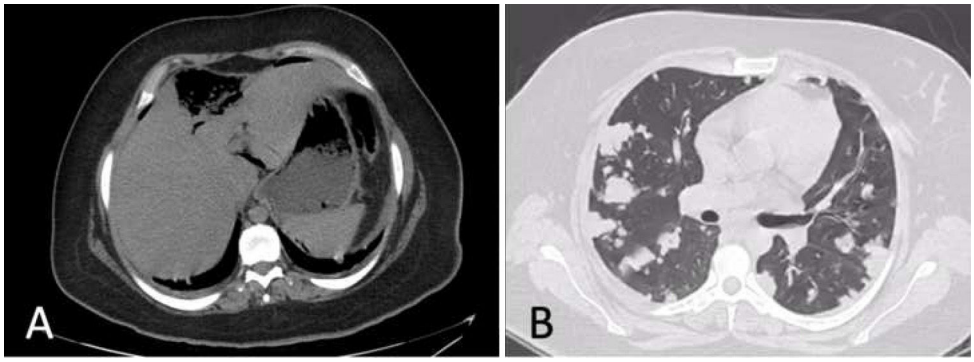

Figure 2 A) Abdominal CT scan showing a ruptured liver abscess. B) Chest CT scan showing multiple bilateral cavitated nodules.

unilocular liver abscess was more virulent than the multilocular disease. Younger patients with a stronger immune system were hypothesized to present with more aggressive clinical symptoms due to findings of a higher APACHE II score, septic shock, metastatic infection, acute respiratory failure, and the formation of gas in the abscess observed in imaging studies[7].

Chou et al.[7] described pulmonary septic embolism, a complication that has not been widely studied, and reported a rate of 4.5% in a total of 14 cases. Eighty-three percent of those cases presented with diabetes mellitus and their prevalent symptoms were fever and dyspnea. Only one patient in their case series had abdominal pain upon hospital admission. In addition, those authors considered that abdominal ultrasound should be ordered to rule out liver abscess in diabetic patients presenting with fever, dyspnea, and a chest x-ray revealing multiple opacities and infiltrative patterns.

It is important to remember that pyogenic liver abscesses account for 75% of liver abscesses in industrialized countries, but only 66% of the bacterial pathogens are identified. In the United States, the increase in frequency of *K. pneumoniae* liver abscesses may reflect an increase in the Asian population seen at their institutions or be due to a previously unreported hypervirulent serotype[8]. To the best of our knowledge, the present case of invasive liver abscess syndrome is the first to be reported in Latin America. This syndrome should be suspected when a patient is immunocompromised and chest x-ray reveals infiltrates, along with an image suggestive of liver abscess, so that he/she can be treated opportunely, preventing the high mortality resulting from invasive liver abscess syndrome.

Ethical disclosures

The authors declare that no experiments were conducted on humans or animals for the present study, that they have followed the protocols of their work center on the publication of patient data, and that they have preserved patient anonymity at all times. Informed consent was not required for the publication of the present case because the article contains no personal data that could identify the patient.

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Conflict of interest

The authors declare that there is no conflict of interest.

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Allgrove syndrome in a toddler: Alacrima and achalasia, with no adrenal insufficiency☆

Síndrome de Allgrove en lactante: alacrimia, acalasia, sin insuficiencia suprarrenal

Allgrove syndrome (AS) is a progressive, neuroendocrine disorder of unknown etiology, with an autosomal recessive inheritance pattern, that is characterized by achalasia, alacrima, adrenal insufficiency, and autonomic and neurologic alterations. It involves homozygous mutations in the AAAS gene that is located on chromosome12q13 and encodes the ALADIN protein (c.1331 + 1G > A is the most widely reported mutation worldwide), and the estimated prevalence is 1/1,000,000 persons[1,2].

A 21-month-old female toddler had a full-term birth history. She was the second child born to healthy nonconsanguineous parents and had a healthy brother. Postnatal weight was 3.075 kg, postnatal length was 48 cm, and she had no perinatal hypoxia. Ever since birth, the patient did not produce tears. She presented with malnutrition (weight gain of 900 g in 18 months), with episodes of vomiting starting at 9 months of age (daily, abundant, and immediately postprandial, consisting of undigested food), and dysphagia to solids (ingesting 65 kcal/kg/day). She received prokinetics, proton pump inhibitors (PPIs), and extensively hydrolyzed formula, with no improvement.

Physical examination revealed weight of 6.2 kg, length 74 cm, head circumference 43 cm, no characteristic facies, partial alopecia, no tears, generalized muscle weakness and atrophy, global developmental delay (GDD), and severe chronic malnutrition. No cutaneous hyperpigmentation was observed.

The patient was admitted to the hospital for nutritional recovery through enteral feeding, in accordance with the World Health Organization guidelines for in-hospital treatment of children with severe malnutrition. A contrast esophagram series showed a ''bird's beak'' appearance (Fig. 1). Upper gastrointestinal endoscopy revealed dilation of the distal esophagus with food remnants. High-resolution esophageal manometry identified type I esophageal achalasia (EA) (Fig. 2). Schirmer's test was positive and the levels of adrenocorticotropic hormone (ACTH) and cortisol were normal.

The patient underwent laparoscopic Heller cardiomyotomy and partial anterior Dor fundoplication, with no complications, and 14 months later, presented with dysphagia to solids. Slight dilation of the distal esophagus was observed, and it was dilated to 10 mm with a through-the-scope pneumatic balloon, at the level of the lower esophageal sphincter (LES).

Currently, at the age of four years and 2 months, the patient has adequate esophageal distensibility, no stricture, and a functional surgical fundoplication. Her nutritional status has improved (mild chronic malnutrition) and her growth velocity was 6.5 cm/year. There is no corneal epithelial defect and no adrenal insufficiency. The patient is receiving physical and occupational therapy due to her GDD and is under treatment with prokinetics and a homemade blenderized diet because of regurgitation.

There are few reports of pediatric cases of this disease in Latin America, and to the best of our knowledge, the present case is the first report of AS in a Latin American toddler. In publications from Mexico, alacrima was reported as the primary symptom, followed by achalasia. Adrenal insufficiency was documented in one of 3 patients, at 5 years of age. Another patient presented with autonomic and peripheral neurologic dysfunction[3–5].

Full-term neonates present with tears from the first day of extrauterine life and lacrimal fluid production is completely developed between 1 and 7 weeks of life. Alacrima is considered an early symptom of AS, and when present, the syndrome should be included in the differential diagnosis[6].

EA is a motility disorder with an estimated annual prevalence of 0.18/100,000 children. Symptoms are vomiting,

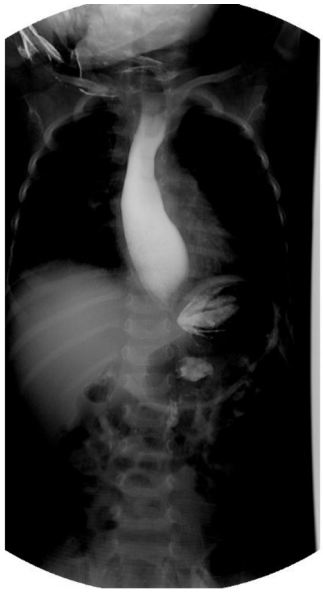

Figure 1 Esophagram showing a dilated esophagus at the level of the esophagogastric junction, with an accentuated narrowing of the distal esophageal lumen and ''bird's beak'' appearance.

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