Abstract:
The presenting manifestations of systemic lupus erythematosus (SLE) are diverse and can present insidiously over years with nonspecific complaints or acutely with life-threatening symptoms. We report a case of a previously healthy 18-year-old female who presented to the pediatric emergency department with altered mental status, multi-system organ failure, and fulminant sepsis from pneumococcal meningitis. Further work-up of this patient confirmed a new diagnosis of SLE. This case highlights the potential degree of immune dysfunction in patients with SLE by opportunistic agents and common pathogens. A high index of suspicion for SLE in childhood is important as delays in diagnosis can lead to life-threatening complications and long-term sequelae.

Keywords:
altered mental status; sepsis; meningitis; pneumococcal infection; renal failure; multi system organ failure; systemic lupus erythematosus

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Adolescent with Abdominal Pain and Altered Mental Status: Are You Confused?

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A previously healthy 18-year-old female was brought to the emergency department (ED) by EMS with a chief complaint of abdominal pain for 2 days. She was in her usual state of health until two days prior when she had onset of abdominal pain with one episode of non-bloody, non-bilious vomiting that coincided with the onset of menstruation. These symptoms were similar to her prior symptoms of dysmenorrhea. However, over the next 2 days, her abdominal symptoms worsened and she also reported a one day history of progressive periorbital swelling, tactile fever, and 3 episodes of non-bloody, loose stools. On the day of presentation, she became confused with slurred speech and was unable to ambulate. En route to the ED, she had an episode of stool and urine incontinence.

On review of systems, there was no recent history of upper respiratory infection, headaches, neck pain or stiffness, joint pain, trauma, or sick contacts. There was no history of recent travel, new foods or medications. She had no known drug allergies. There was no history of sexual activity or tampon use.

Her past medical history was notable only for dysmenorrhea. The patient lived with her mother, 4 siblings and grandmother. The family medical history was significant for a maternal history of asthma and a father who died in his 30's from end-stage renal disease.

On physical examination, the patient was alert, oriented to time, place and person but had intermittent episodes of confusion. Her initial vital signs included: temperature 102.9 °F orally, heart rate 34 beats/min, respiratory rate 24/min, blood pressure 119/80 mm Hg, and her oxygen saturation was 100% on room air. Her weight was 64.5 kg. Her HEENT exam was significant for non-tender, bilateral mild periorbital swelling without any signs of trauma or erythema. Her pupils were symmetric, round and equally reactive to light. Extraocular movements were intact. Her neck was supple, without pain or stiffness with range of motion. Her breath sounds were symmetric and clear bilaterally. Cardiac exam revealed tachycardia with normal S1 and S2 without murmur. Her abdomen was soft, non-distended but tender to palpation of the lower quadrants bilaterally, with no rebound tenderness or guarding. She had no abdominal masses and her liver edge was palpable just below the right costal margin. Her lower legs had 1+ pitting edema bilaterally. Her skin was dry, warm and well perfused with scattered hyperpigmented macules on her arms and legs bilaterally. There was no bruising, petechiae or purpura. Her confusion made it difficult to assess her ability to follow commands, however, she was able to move all her extremities and had 2+ deep tendon reflexes.

Initial laboratory results showed a white blood cell count of 10,600/mm^3 (normal 4,500 – 12,000 mm^3) with a differential of 85% neutrophils, 9% lymphocytes, 4% monocytes and 0% eosinophils. The hemoglobin was low at 5.2 g/dL (12.3-15.3 g/dL), the platelet count was 154,000/mm^3 (150,000-400,000/mm^3) and the reticulocyte count was 2.6% (0.8-2.2%). Serum electrolytes were significant for a bicarbonate of 17 mEq/L (22-26 mEq/L), blood urea nitrogen of 42 mg/dL (6-20 mg/dL) and creatinine of 1.8 mg/dL (0.5-1mg/dL), with normal values for serum sodium, potassium, chloride, glucose, calcium, magnesium and phosphorus. Liver function tests revealed an albumin of 1.5 g/dL (3.1-5.4 g/dL), a total protein 6.3 g/dL (6-8 g/dL), and with remaining values all normal. The erythrocyte sedimentation rate was 140 mm/h (< 21 mm/h). Her urinalysis results included a specific gravity of >1.030, 300 mg/dL protein, and large blood on dipstick. Blood, stool, and urine cultures were sent. Serum toxicology and urine drug screens were negative. Chest radiograph demonstrated diffuse interstitial infiltrates, bibasilar consolidations, with a left pleural effusion. Computed tomography of the head was negative for mass lesions, intracranial bleed, or enhancing lesions, but showed pansinusitis. Cerebrospinal fluid analysis showed 3 WBC/mm^3 (0-7/mm^3), 8 RBC/mm^3, glucose of 62 mg/dL (40-80 mg/dL) and protein of 13 mg/dL (5-40 mg/dL). Gram stain of the cerebrospinal fluid showed gram positive cocci from the first tube but was negative on the fourth tube.

While in the ED, she continued to have multiple episodes of loose stool and became increasingly confused and agitated. She was also oliguric despite attempts at fluid resuscitation with normal saline, 5% albumin and a packed red blood cell transfusion. She was given one dose of ceftriaxone intravenously. Pediatric nephrology, hematology/oncology and infectious disease services were consulted. The patient was admitted to the intensive care unit for further management. On hospital day 2, a diagnosis was made.

In summary, this is a complicated picture of a previously healthy 18-year-old female who presents to the ED with altered mental status associated with fever, peripheral edema, and a hyperpigmented macular rash. Her initial evaluation showed severe anemia, hypoalbuminemia, elevated inflammatory markers, pleural effusion and renal failure. This constellation of findings suggests a multi-organ system disease process.

DIFFERENTIAL DIAGNOSIS

The patient who presents to the ED with altered mental status is always a cause for alarm, and requires careful scrutiny. The differential diagnosis of altered mental status is extensive and lends itself to a wide range of effective, life-saving management options if there is prompt recognition of the etiology. A complete list of conditions that present as altered mental status would not be practical. Therefore, only the most common etiologies relevant to our case discussion will be presented here for review.

One approach to altered mental status is to divide the etiologies into 2 groups: structural and medical (Table 1). In general, structural causes often have focal neurologic findings with pupillary abnormality (asymmetric and/or non-reactive), while medical causes usually result in non-focal, generalized neurologic dysfunction, often with normal pupillary findings, except for toxicologic etiologies. A focused history and physical examination to rapidly assess the level of consciousness, pupillary response, extraocular movements, and motor response to pain is an essential component of the initial evaluation and may direct appropriate treatment.[1]

Structural causes of altered mental status include traumatic injuries, tumors, and vascular lesions. Traumatic injuries can lead to shearing injuries, which can cause brain contusions and rupture of blood vessels leading to intracranial bleeding. These