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Infected cystic hygroma resulting in septic shock and respiratory failure: A case report

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ABSTRACT

Macrocystic lymphatic malformations (cystic hygromas) are a common cause of cystic neck lesions. These lesions are often diagnosed prenatally in children. In cases without airway compromise, these children are discharged from the hospital for elective treatment. Surgical excision is one treatment modality while sclerotherapy has recently shown adequate results as well. While infection is a relatively common problem for lymphatic malformations, the majority can be treated with antibiotics alone. We present a case in which septic shock and respiratory failure resulted from primary infection of a macrocystic lymphatic malformation in a term infant discharged with a lymphatic malformation of the neck. Urgent surgical drainage failed, and complete excision was ultimately required for source control due to numerous small multiloculated small cysts inaccessible via incision and drainage.

Macrocystic lymphatic malformations (formerly termed "cystic hygromas") occur in nearly 1:2000 live-born children and are most commonly located in the head and neck region [1–3]. With improvements in ultrasound imaging, these lymphatic malformations are often seen as early as the first trimester [4]. Surgical excision is generally regarded as definitive treatment, however, sclerotherapy has been advocated for uncomplicated macrocystic lymphatic malformations without evidence of airway compromise [5,6]. Several novel therapies have recently been proposed including use of sildenafil, propranolol, and sirolimus and have shown varying success in small case reports and case series [7–9]. Reports of spontaneous regression of lymphatic malformations have also been reported although rare [5]. A relatively common complication of macrocystic lymphatic malformation is infection which may often be treated successfully with antibiotics alone followed by elective resection or sclerotherapy. We present a case of a large cervical lymphatic malformation in a neonate who developed septic shock and respiratory failure prior to treatment. The child also failed conservative attempts at drainage, and urgent complete surgical excision was required for source control. The operative findings explaining failure of medical treatment and suggestions for definitive management of severe infection in this situation are reviewed.

1. Case report

A 6-day-old male born at term with history of intrauterine methamphetamine exposure and prenatally diagnosed cervical macrocystic lymphatic malformation presented to the emergency department (ED) with poor feeding and an increased work of breathing associated with enlargement and discoloration of the lymphangioma. Examination revealed sternal retractions, tachypnea, hypoxia on room air, tachycardia, and a large 10 × 10 cm cystic mass on the right neck with new, small areas of erythema and ecchymosis (Fig. 1). Laboratory testing revealed severe leukopenia and lymphocytopenia, indirect hyperbilirubinemia, and elevated coagulation tests (PT, PTT); urinalysis was negative for infection. Chest X-ray (CXR) performed was without evidence of tracheal deviation and compression. Sepsis workup commenced and the child was admitted to the Neonatal Intensive Care Unit (NICU) for worsening respiratory distress and placed on empiric ampicillin and gentamicin. The patient's lymphangioma remained soft and the patient never developed any signs of tracheal compromise or carbon dioxide retention. Endotracheal intubation was performed for worsening respiratory distress without difficulty and an adequate air leak around the endotracheal tube suggested the airway was not compromised. Clinically, the respiratory distress, hemodynamic

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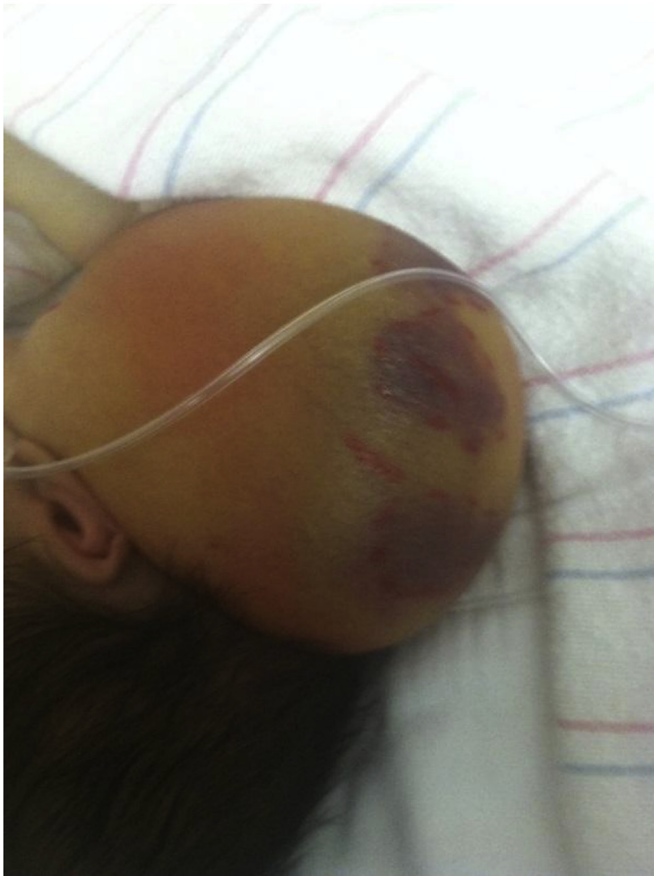

Fig. 1. The child's macrocystic lymphangioma at the time of admission showing new onset erythema and areas of ecchymosis.

compromise and rapid decline appeared to be from overwhelming sepsis. Over the next few hours oxygenation became more difficult and high-frequency oscillator ventilation was required. Blood cultures became positive at 5 h for gram-negative bacteremia which ultimately speciated into *Escherichia coli* resistant to ampicillin and fluoroquinolones. Surgical consultation was obtained on the morning of admission for the inflamed and necrotic-appearing neck lymphangioma (Fig. 2). Urgent bedside needle decompression was performed immediately expressing purulent fluid.

The patient continued to decline in status throughout that day and the decision was made to perform bedside incision and drainage with placement of a Penrose drain (Fig. 3). Care was made to break up the septations in the wound at the time of drainage.

The patient continued intravenous (IV) antibiotic therapy for three more days but necrosis of the wound progressed and the patient again decompensated over 24 h requiring increased vasopressor and ventilatory support. Due to the patient's critical instability secondary to sepsis from the infected lymphatic malformation, and failure of more conservative attempts, it was felt that the only option for source control was complete surgical excision. The necrotic wound was excised with electrocautery and the spinal accessory nerve was identified and isolated. Several very small, pus-filled lymphatic cysts had remained which contained frank pus, and partial necrosis of the lymphangioma was noted to extend to the base of the malformation (Fig. 4). Complete excision was performed and the margins were locally advanced to provide a loose, but adequate wound closure over a Penrose drain.

After the procedure, the child showed rapid improvement over the next several hours and was weaned off vasopressors and to

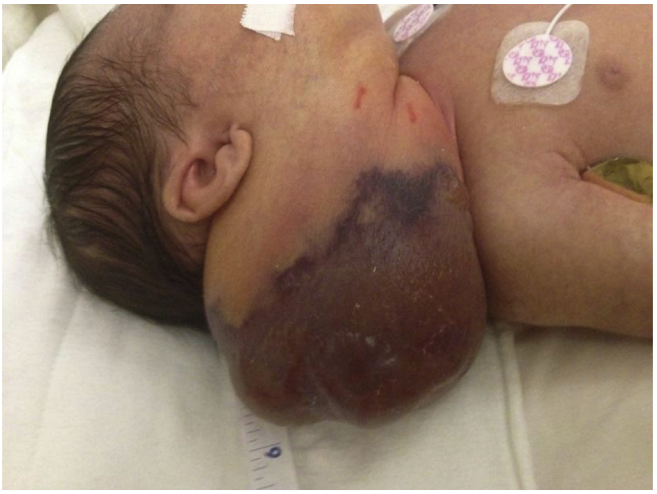

Fig. 2. The infant has been intubated for respiratory failure. The lymphangioma has enlarged and shows bullae and areas of necrosis.

conventional mechanical ventilation. The drain was removed after 2 days and the wound was loosely packed with calcium alginate. On postoperative day three, the patient was extubated after remaining a day on minimal ventilator settings and exhibiting a clinical air leak. He remained on intravenous cefotaxime for a total of fourteen days after postoperative cultures and sensitivities were obtained and was ultimately discharged on postoperative day 15. Upon return to the surgical clinic, he was noted to have torticollis which resolved after several weeks of physical therapy and had a favorable cosmetic result (Fig. 5) without evidence of a winged scapula or any nerve damage.

2. Discussion

To our knowledge, this is the first reported case of septic shock and respiratory failure resulting from primary infection of a cervical lymphatic malformation. Conservative management of the infection with systemic antibiotics in this case was initially attempted but did not result in improvement. Neither needle decompression nor incision and drainage of the infected cyst were adequate to stem the clinical deterioration, and the patient progressed to

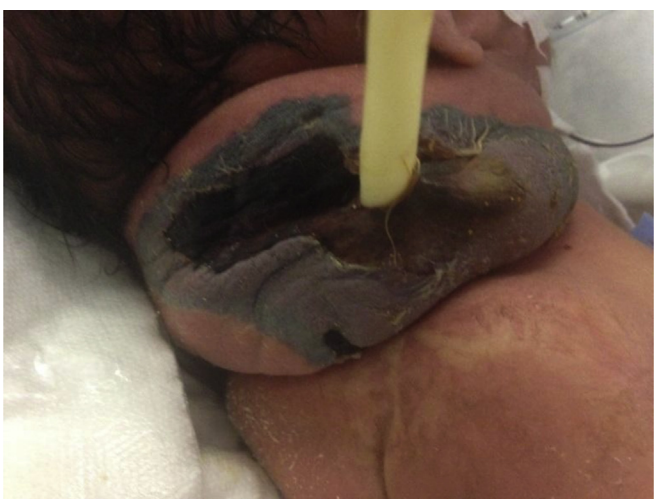

Fig. 3. The infant has developed systemic sepsis and skin necrosis, despite incision and drainage with care to break up septations.