Bridge to diagnosis: The use of extracorporeal membrane oxygenation in a child with interstitial lung disease secondary to clinically amyopathic dermatomyositis

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Clinically amyopathic dermatomyositis (CADM), a subtype of dermatomyositis (DM), was first described by Sontheimer in 2002, and is characterized as having the classic cutaneous manifestations of DM but with little or no evidence of myositis [1]. The presence of anti-melanoma differentiation associated gene 5 (anti-MDA-5) autoantibodies, in the setting of CADM, is associated with rapidly progressive interstitial lung disease (RPILD) and has a poor prognosis with high rates of mortality [2]. The use of extracorporeal membrane oxygenation (ECMO) in patients with RPILD has been well described in adults, but not in children. Herein we present a 14-year old patient with interstitial lung disease, initially of unknown etiology, whom we supported with ECMO in the acute phase of her illness for nearly two months during which the diagnosis of anti-MDA-5 dermatomyositis was established.

1. Case report

A 14-year old female patient, with a 3 month history of progressively worsening constitutional symptoms including dyspnea, fever, and a 35-pound weight loss, was transferred to our institution for an ECMO evaluation after developing acute respiratory distress syndrome (ARDS) requiring intubation, mechanical ventilation, and chest tube placement for bilateral pneumothoraces. Her past medical history included mild intermittent asthma, but was otherwise unremarkable. Chest radiograph at our institution demonstrated dense consolidations bilaterally consistent with pneumonia (Fig 1). Computerized tomography (CT) scan of the chest showed diffuse patchy bilateral airspace disease, extensive pneumomediastinum and subcutaneous emphysema (Fig 2). Flexible bronchoscopy revealed diffuse blood in the small airways consistent with the evolution of ARDS, and a lung biopsy showed organizing, diffuse alveolar damage with hyaline membrane and fibrin plug formation. She was hypoxemic and unable to be adequately ventilated on conventional ventilator. Thus, high frequency oscillatory ventilation (HFOV) with inhaled nitric oxide was initiated. Despite the increased support, her respiratory status worsened and on hospital day 13, veno-venous ECMO (V–V ECMO)
dermatomyositis with presence of anti-MDA-5 autoantibodies. However, her presentation was atypical as there was no evidence of cutaneous involvement or vasculopathy. On hospital day 41, she was started on immunosuppressive therapy including pulse steroids, intravenous immunoglobulin (IVIG), tacrolimus, and rituximab. In addition, she received 5 rounds of plasmapheresis. Her family deferred muscle biopsy, but ultrasound of the left deltoid showed no evidence of myositis. During immunosuppressive therapy, while on ECMO, her respiratory and cardiac status slowly improved. She was eventually weaned from high frequency oscillation to a conventional ventilator, and on hospital 69, she was decannulated from ECMO after a 55-day total run.

Three weeks after ECMO support was discontinued, the patient’s clinical status abruptly deteriorated. She developed aspiration pneumonia and went into septic shock and acute renal failure. Blood cultures at that time were positive for acinetobacter baumannii. Broad-spectrum antibiotics were subsequently initiated. She was placed back on HFOV and inotropes were started. Echocardiogram showed severely diminished right ventricular function. Seizure activity was also noted and an electroencephalogram (EEG) showed epileptic potential in the left frontal-temporal region and severe slowing. She had absent brain stem reflexes on exam. After an inter-disciplinary family meeting, care was withdrawn and the patient expired. Family refused autopsy.

2. Discussion

CADM-associated interstitial lung disease, in the setting of anti-MDA-5 antibodies, is a life threatening complication of dermatomyositis with mortality rates nearing 60%. The difficulty in establishing the diagnosis of CADM often leads to a delay in therapy. The detection of anti-MDA-5 autoantibodies is useful in predicting and monitoring response to treatment. In our patient with ARDS, CADM was not initially considered as her presentation was atypical. There was no evidence of cutaneous involvement or a history of myositis that is normally associated with classic CADM. To complicate matters, her clinical course deteriorated rapidly and confounded her diagnosis. Initiating ECMO to treat her ARDS, and ultimately serve as a bridge to her diagnosis, was critical. The mortality rate of pediatric patients with ARDS, who have become refractory to conventional respiratory ventilation, exceeds 90%. With ECMO, however, survival to hospital discharge for pediatric patients with severe respiratory failure is 56%, [4]. Our early management strategy was to support the patient with ECMO therapy until a diagnosis was established.

Given the acute onset and decompensation of our patient, with respect to her age and lack of previous significant medical history, ECMO was instituted immediately once she met institutional criteria. Once ECMO was instituted, the palliative care team was consulted to help mediate ethical conversations between the intensivists, medical subspecialists and the patient’s family. The collaborative medical team thoroughly discussed the use of ECMO as a bridge to diagnosis with our patient’s parents. It was the parents wish to support their daughter on ECMO as long as necessary to establish a diagnosis provided she was not decompensating further or suffering in pain. The medical team agreed with this plan.

The presence of anti-MDA-5 autoantibodies was detected through enzyme-linked immunosorbent assay (ELISA) testing 28 days after we instituted ECMO and approximately 4 months after her initial symptoms. She remained on ECMO, undergoing intense immunosuppressive therapy, for an additional 27 days before we discontinued ECMO for a total run of 55-days.

Outcomes are generally favorable with short term ECMO therapy, but prolonged ECMO therapy in children with refractory
pulmonary failure is associated with low survival and significant long-term sequelae. [5]. Although our patient ultimately succumbed to her illness, her prolonged survival on ECMO was the result of a proactive and multidisciplinary approach from the ECMO service, the intensivists, and all consulting services.

References


