Case 9

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Clinical Abstract:

The patient was a 5 year old girl who presented 3 weeks ante mortem with facial swelling and total body weakness. The swelling progressed to involve the upper then lower extremities. She was admitted to a local clinic for evaluation. An urine culture was positive for Staphyloccocus aureus and she was treated for pyelonephritis with several antibiotics including gentamicin and nitrofurantoin. Her past medical history was unremarkable, however, her vaccinations status was not known. Her parents and 2 year old sister had been in good health.

Despite antibiotic therapy, her condition continued to deteriorate over the next 2 weeks until she could no longer walk or hold herself upright. Five days later she was admitted to Children's hospital for further evaluation. She was initially cooperative yet lethargic. Her temperature was 100° F with a pulse rate of 102, respiratory rate of 24 and blood pressure of 100/52. There was some soft tissue edema in the thighs and periorbital tissues. Muscle tenderness was limited to the gastrocnemius. Neurologic evaluation revealed virtually no function of her proximal extremities with 1/4+ and 2/4+ function in her distal upper and lower extremities, respectively. Deep tendon reflexes were 1+ bilaterally in both the upper and lower extremities. Sensory, cranial nerve, and ophthamalogic examinations were normal. No rash was noted on her skin. Respiratory and cardiovacular examinations were unrevealing. Laboratory studies demonstrated a CK of 16,804 U/L (95% MM, 5% MB); SGOT 1085; SGPT of 481; ANA of 1:640; normal electrolytes and pseudocholincasterase levels. CSF was normal. The following day she suffered multiple cardiac arrests. Pericardicentesis recovered 30 mls. of straw colored fluid. After 2 hours of resuscitation, no cardiac contractility was demonstrated by ultrasound and resuscitative efforts were stopped.

The autopsy was limited to the chest, abdomen, and extremities. The findings included serous pleural effusions, bilateral lower lobe infiltrates, and serous pericardial effusions. Cardiac and skeletal muscles were diffusely edematous. The kidneys, lungs,and other organ systems were slightly swollen yet histologically normal. Bacterial, fungal, and viral cultures of the liver and heart were negative.

Materials submitted: One H & E slide from skeletal muscle

One 2 X 2 Kodachrome of electron micrograph from skeletal muscle endothelial cell.

Points for discussion: Diagnosis
Pathogenesis