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Case report

A fatal presentation of dermatomyositis with facial swelling

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ABSTRACT

Juvenile dermatomyositis (JDM) is the most common inflammatory autoimmune myopathy in children. Most common presentations consist of heliotrophic rash and/or gottron's papules in addition to proximal muscle weakness. A typical presentations have been reported. We present a 13-year-old African American male who presented with a two-week history of bilateral periorbital edema that was unresponsive to glucocorticoids. He had elevated transaminases but no detectable muscle weakness. A muscle biopsy was consistent with juvenile dermatomyositis. This case highlights the need to consider dermatomyositis in cases of facial swelling and the use of aggressive immunosuppressive therapies due to its associated vasculopathies.

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1. Case report

A 13-year-old African American male presented with a two-week history of bilateral periorbital swelling, decreased appetite and fatigue. Because of a history of both eczema and allergies to pollens, aspirin, and peanuts, he was treated with anti-histamines and topical steroids. When the swelling did not improve, a CT scan of his sinuses was performed that showed inflammation lining the maxillary, ethmoid and sphenoid sinuses. He was treated with azithromycin (Z-Pak) and Omnicef. Although there was a history of subjective fever, there was no history of muscle weakness, oral ulcers, shortness of breath, itching, skin tightening or recent immunizations. He was an active athlete, played football and lifted weights (up to 80 pounds) regularly. When the periorbital swelling did not improve after treatment with antibiotics, he was admitted to the hospital for further evaluation.

At the time of admission, he had bilateral periorbital swelling without erythema or rash (Fig. 1, panel I). A dried yellow pus-like discharge was present in the left eye. Although he complained of blurry vision, he was able to open his right eye with difficulty. The only other abnormality was a dry eczematous rash over both anticubital fossae. His muscle strength tested as 5/5 in all four extremities for both proximal and distal muscle groups.

Laboratory values and infectious disease studies are shown in Tables 1 and 2. The abnormalities included a low white blood count (2.6×10^3/uL), elevated transaminases [aspartate aminotransferase (AST) 292 and alanine aminotransferase (ALT) 85], elevated creatine kinase of 5037 and an elevated aldolase of 28.2. The patient was started on high dose glucocorticoids and was discharged from the hospital.

Despite treatment, the patient did not improve. One month later, he developed upper and lower lip edema (Fig. 1, panel II) and an intermittent low grade fever. The periorbital edema persisted and he acquired an erythematous rash covering his chest and upper arms together with diffuse swelling in both upper arms. Muscle weakness in the hip flexors and shoulder abductors was detected for the first time, eight weeks after the initial presentation. An enlarged left supraclavicular lymph node was also noted. A node biopsy and bone marrow evaluation were performed and neither test gave evidence of a malignancy, although the WBC count decreased to 2.1 and the transaminase enzymes increased (AST, 465; ALT, 99). A trial of systemic anti-fungal medications and acyclovir was begun and a muscle biopsy was performed.

2. Final diagnosis

The biopsy of the sternocleidomastoid muscle showed the presence of inflammatory cells along with perifascicular necrosis

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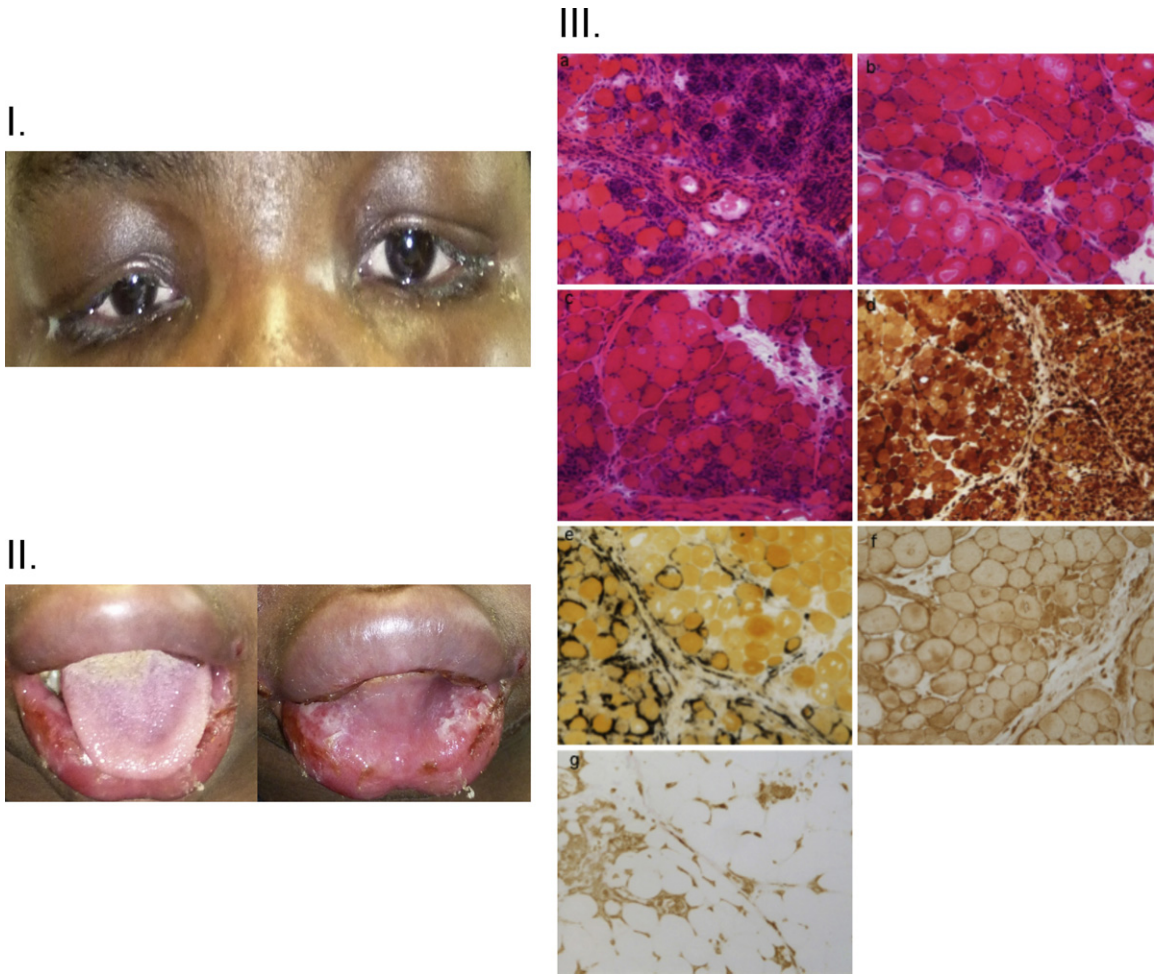

Fig. 1. Panel (I) *Periorbital swelling*. Periorbital edema at initial presentation; panel (II) *Lip swelling*. The left and right panels indicate the degree of lip swelling noted one month after initial presentation; panel (III) *Muscle biopsy*. (a) Foci of severe muscle necrosis with phagocytosis; (b and c) perifascicular necrosis and vacuolization of muscle fibers; (d) non specific esterase stain showing dark staining macrophages invading necrotic fibers and in the interstitial area (× 40); (e) Alkaline phosphatase stain showing increase activity in capillaries and vessels, (× 100); (f) Inmunoperoxidase stain for MHC-1 showing upregulation of MHC-1 in the surface of muscle fibers; (g) Inmunoperoxidase for CD45 showing numerous positive staining lymphocytes (× 100).

suggesting severe active necrotizing myopathy, consistent with juvenile dermatomyositis (JDM) (Fig. 1, panel III).

3. Hospital course

Later that evening, the patient developed bloody diarrhea, which led rapidly to metabolic acidosis, hyperkalemia, and ventricular fibrillation. Despite resuscitative measures and replacement fluids, he succumbed the following day. A final autopsy revealed two large ulcers in the duodenum with erosion into a submucosal blood vessel. A massive amount of blood was found in the peritoneal cavity indicating intestinal perforation as a cause of hemorrhage and death. Examination of the skeletal muscles identified myonecrosis with inflammation consistent with

Table 1
Laboratory values.

	Normal range	At presentation	Eight weeks	Nine weeks
Sodium	134–143 mmol/L	136	124	117
Potassium	3.5–5.0 mmol/L	4.6	4.1	4.8
Chloride	96–109 mmol/L	99	92	89
CO_2	20–31 mmol/L	28	26	25
Urea nitrogen	7–17 mg/dL	10	12	23
Serum glucose	60–115 mg/dL	103	96	104
Calcium	8.8–10.8 mg/dL	9	8.3	7.3
Creatinine	0.20–0.70 mg/dL	0.77	0.76	0.76
Total protein	6.3–8.2 g/dL	8.1	6.4	5.2
Albumin	3.5–5.4 g/dL	4.4	3.5	2.5
Total bilirubin	0.2–1.2 mg/dL	0.4	0.5	0.5
Lactate dehydrogenase	470–750 units/L	1922	4537	–
Alkaline phosphatase	178–455 units/L	147	77	105
Aspartate aminotransferase	15–40 units/L	292	791	1421
Alanine aminotransferase	10–55 units/L	85	170	193