

Periorbital and facial swelling and fever

Sunday, 01 October 2006

A 55-year-old female patient presented to us with a 10-day history of malaise, lethargy, and fever up to 39.50 C accompanied by rigors. She also complained of diffuse myalgias, arthralgias, and bilateral facial edema. Her past medical history was significant for a modified mastectomy for invasive breast cancer in 2000 but she is free of disease 6 years later. There was no history of trauma. She admitted that she had botulinum toxin injections in the forehead 4 years ago but there was no prosthesis in the periorbital area (neither silicone nor fat).

On examination, her blood pressure was 130/70 mm Hg and she was feverish (39.0 C). Pulse rate was 90 bpm regular and respirations were 18/ minute. There was marked edema of the face, especially in the periorbital and malar areas (Figure 1), and this was accompanied by erythema and sensitivity. There was no associated feeling of pruritus of the aforementioned area. A scar from the previous surgical operation in the breast area was evident.

Laboratory examinations showed leucocytosis (12480 /mm³ with a left shift), normal eosinophil count, increased C-reactive protein [11.4 mg/dl (normal values 0-0.5)], and increased erythrocyte sedimentation rate [70 mm/1st hour (normal 0-20)]. Creatinophosphokinase levels were increased [1453 IU/l (normal 21-215)]. Immunoglobulin levels were increased for IgA [550 (normal 55-377) mg/dl] and IgE [(696 IU/l (normal 0-200))], Serum protein electrophoresis was normal. C1 esterase inhibitor levels were normal [339 mg/l (normal 210-390)]. Antinuclear autoantibodies, anti-dsDNA and anti-Sm were all negative. Serology testing was negative for *Trichinella spiralis* and *C. trachomatis*. Magnetic resonance angiography of the brain vessels was normal. MRI imaging confirmed the presence of edema in the periorbital area. Electromyography of the muscles of the upper and lower limbs was normal. What is the diagnosis?

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Diagnosis

Biopsy of the muscles of the periorbital area revealed dermatomyositis. Anti- Jo1 auto-antibodies were negative. CT of the neck, thorax and the abdomen was normal as was bone scintigraphy. Upper gastrointestinal endoscopy was normal. At colonoscopy, a 1 cm sessile polyp was resected; pathologic examination showed that this was hyperplastic without evidence of malignancy.

Initial administration of antibiotics to our patient had no effect. Once biopsy results were available steroids led to marked clinical improvement and laboratory indices returned to normal. When seen for follow up the patient was in excellent health. Despite the improvement of our patient, she is also under close follow up of the oncology team.

Teaching points

- Bilateral periorbital edema in dermatomyositis is an occasionally reported sign of the disease. The dusky purple heliotrope rash is usually present. It may be the only sign of the disease. Sometimes no other anatomical region except for the periorbital area is afflicted, at least in the beginning of the disease.¹ Other skin pathology includes Gottron's papules, cuticular erythema, and telangiectasia, papulosquamous eruption of the hairline, face and trunk. A vesicobullous form of dermatomyositis is a rare manifestation. Fever may also be a presenting sign of the disease. Diseases leading to decrease levels of serum albumin such as cirrhosis, nephrotic syndrome, and protein-losing enteropathy lead usually to generalized edema in addition to the periorbital area, but usually cause no fever. Allergic diseases are other common causes of periorbital swelling usually not accompanied by fever. Other diseases that enter the differential diagnosis are trichinosis, systemic lupus erythematosus, TNF-receptor associated periodic syndrome, and bilateral

internal jugular thrombosis.2-4

- The diagnosis of dermatomyositis may evade if the extent of the disease is limited. Corticosteroids are the mainstay of the treatment of dermatomyositis. The association of dermatomyositis, especially in patients over 40, with neoplasia is well-known.⁵ Ruling out an underlying malignancy especially when a previous history of the disease is present is mandatory.

Acknowledgement

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References

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