Clinical Vistas

Periorbital and facial swelling due to dermatomyositis

A 55-year-old woman presented with a 10-day history of malaise, lethargy and an elevated temperature (up to 39.5°C) accompanied by rigor. She also reported diffuse myalgia, arthralgia and bilateral facial edema. The patient's medical history included a modified mastectomy for invasive breast cancer 6 years earlier, but at the time of presentation she was free of malignant disease. There was no history of trauma. Botulinum toxin had been injected into her forehead 4 years before presentation, but there had been no cosmetic injection of silicone or fat in the face or periorbital area.

On examination, the patient's blood pressure was 130/70 mm Hg, and she had a fever (temperature 39°C). Her pulse rate was 90 beats/min and regular, and her respiration rate was 18 breaths/min. There was marked edema of her face, especially in the periorbital and malar areas (Fig. 1); this was accompanied by erythema and sensitivity. The patient reported having no associated pruritus. A scar from the mastectomy was evident in the patient's breast area.

Laboratory test results showed leukocytosis (leukocyte count 12.5 × 10^9/L, with a left shift), a normal eosinophil count, an elevated C-reactive protein level (114 [normal 0–5] mg/L) and an increased erythrocyte sedimentation rate (70 [normal 0–20] mm/h). The creatine kinase level was elevated (1453 [normal 21–215] U/L), as were the levels of immunoglobulins IgA (5.50 [normal 0.55–3.77] g/L) and IgE (1670 [normal 0–240] µg/L). Serum protein electrophoresis results were normal, as was the C1 esterase inhibitor level. Test results were negative for antinuclear autoantibodies (anti-dsDNA and anti-Sm) and for *Trichinella spiralis* and *Chlamydia trachomatis*. The results of magnetic resonance angiography of brain vessels were normal. An MRI image confirmed the presence of edema in the periorbital area. Electromyography of the muscles of the upper and lower limbs revealed normal results.

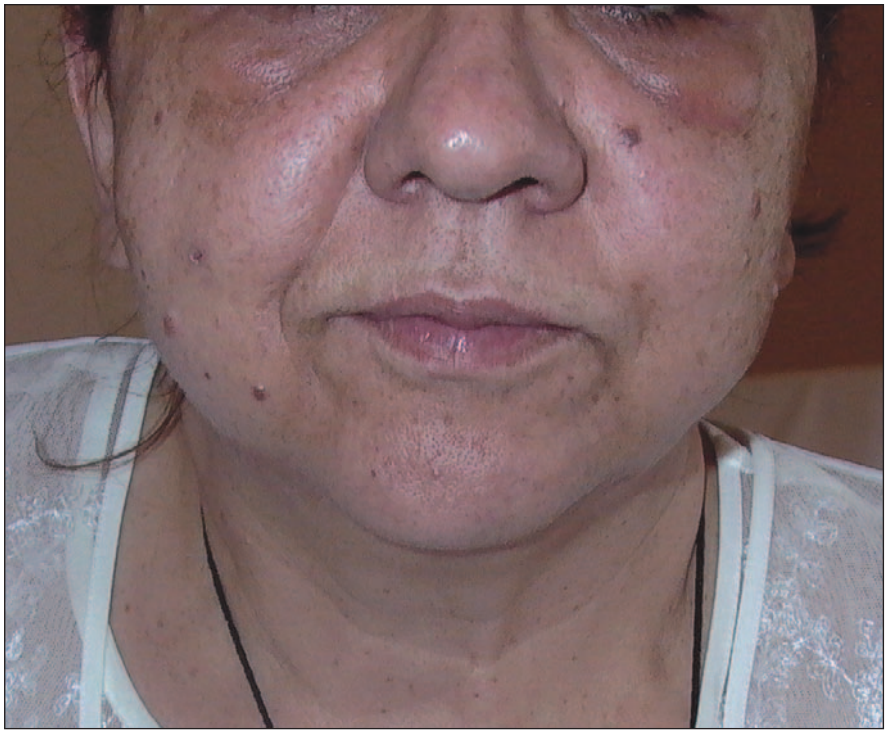

Fig. 1: Facial swelling as the first manifestation of dermatomyositis.

Biopsy of the muscles of the periorbital area revealed dermatomyositis. Test results for anti-Jo1 autoantibodies were negative. CT scans of the neck, thorax and abdomen were normal, as were the results of a bone scintigram. Results of an upper gastrointestinal endoscopy were normal. Colonoscopy revealed a 1-cm sessile polyp; pathologic examination showed that it was hyperplastic but not malignant.

Initial antibiotic therapy had no effect on the edema. After the biopsy results became available, steroid therapy was administered, which led to marked improvement of clinical and laboratory findings. The patient was well at a follow-up examination. Despite the improvement, she is being closely followed by internal medicine and oncology teams.

Dermatomyositis is an autoimmune disease in which the skin and skeletal muscles are inflitrated predominantly by lymphocytes. Although idiopathic, the disease may be the result of an interaction between the patient's genetic background (determining the immune system response) and a viral pathogen. This interaction may lead to activation of the complement cascade and upregulation of the expression of cytokines and chemokines, which results in microangiopathy.[1] Muscle involvement, either diffuse or focal, is a common feature of polymyositis (the skin is not affected). Cutaneous manifestations of dermatomyositis are varied; a localized lilac-coloured (heliotrope) rash around the eyes, nose and cheeks is common. Lesions are also common on the forehead, chest, elbows, knees and knuckles. The exanthem may be pruritic. Bilateral periorbital edema, similar to the presentation by our patient, may occur in acute cases.[2] Particularly in the early stages of the disease, the periorbital area may be the only area affected. Other skin manifestations include Gottron's papules, cuticular erythema, telangiectasia and papulosquamous eruptions on the hairline, face and trunk. A vesico-bullous form of dermatomyositis is rare. Fever may also be a presenting symptom of dermatomyositis. The disease affects twice as many women as men. Laboratory findings associated with dermatomyositis include elevated serum levels of

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skeletal muscle enzymes (e.g., creatine kinase, lactic dehydrogenase and aldehyde-lyase). Electromyography reveals myopathic findings, and skin and muscle biopsies confirm the diagnosis.

The differential diagnosis of bilateral periorbital edema includes trichinosis, systemic lupus erythematosus, periodic syndrome associated with tumour necrosis factor (TNF) receptor, and bilateral internal jugular thrombosis.[3–5] Diseases that lead to decreased levels of serum albumin (e.g., cirrhosis, nephrotic syndrome, protein-losing enteropathy) usually result in generalized edema but not fever. Allergic reactions can also cause periorbital swelling, but are not usually accompanied by fever.

Dermatomyositis may be difficult to diagnose if the extent of the disease is limited. Corticosteroids are the mainstay of treatment, but intravenous immune globulin therapy may also be necessary.[6] Dermatomyositis may be associated with infection (e.g., Lyme disease or enterovirus infection),[7] autoimmune diseases (e.g., rheumatoid arthritis, systemic lupus erythematosus or systemic sclerosis), sarcoidosis[8] and malignant disease. The association of dermatomyositis with neoplasia, especially in patients over 40, is well known. The malignant neoplasms most commonly associated with dermatomyositis are that of the ovary, lung, gastrointestinal tract (pancreatic, colorectal, gastric), and non-Hodgkin's lymphoma.[9] Ruling out an underlying neoplasm, especially if the patient has a history of cancer, is essential.

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