Denouement and Discussion

Juvenile Dermatomyositis

On examination, she had Gottron papules over the extensor surface of metacarpophalangeal and interphalangeal joints (Figure 1), erythematous lichenified plaques on the extensor surface of the elbows and knees (Figure 2), moderate discoloration on the superior palpebral fissure (heliotrope rash). The strength of the upper and lower proximal muscles was 3/5.

Laboratory test results showed an increase in the levels of aspartate aminotransferase, alanine aminotransferase, and creatine kinase. Magnetic resonance imaging (MRI) of the thighs revealed edema and inflammation in almost all of the imaged musculature. An electromyogram showed a myopathic process.

This patient was diagnosed with juvenile dermatomyositis (JDM), an idiopathic, autoimmune vasculopathy that involves the muscle and skin. The incidence is 2 to 3 children per million per year. The most frequent clinical manifestations of JDM are Gottron papules, heliotrope rash, malar rash, and proximal muscle weakness. Nail-fold capillary changes, if present, are important markers of disease activity, and they can be visualized by the unaided eye or with a magnifying tool like dermoscopy (Figure 3).[1] Giant/enlarged and torturous capillaries, loss of capillary areas (dropout areas), and hemorrhages are usually seen in dermatomyositis and scleroderma.[2]

Erythematous or violaceous, scaly, lichenified skin may also be seen on the extensor surface of the knees, elbows (Figure 2), trunk, and extremities. Mucocutaneous ulcers, limb edema, calcinosis, and lipodystrophy are less common skin findings. Muscular involvement includes weakness and loss of function of the proximal muscle groups and anterior neck flexor. Striated muscles of the respiratory or gastrointestinal system may also be affected. The patients may complain of muscle stiffness and tenderness. In chronic cases, restrictive pulmonary disease may ensue. Other symptoms include fatigue, lethargy, arthralgia, and dysphagia.

The diagnosis is based on the combination of the skin findings and myopathy. Bohan and Peter[3] suggested criteria to help establish the diagnosis: characteristic skin rash, symmetric proximal muscle weakness, elevated muscle enzyme levels (creatine kinase, aspartate aminotransferase, lactate dehydrogenase, and aldolase), and confirmation with either a characteristic electromyography or a muscle biopsy.

Our patient had an extensive workup, which included chest radiography, a gastrointestinal series, a bone densitometry test, and a pulmonary function study. All of the results were within the normal limits. In addition, she had an ophthalmologic consultation to rule out signs of intraocular inflammation and to obtain results of a baseline examination prior to treatment with hydroxychloroquine.

Once diagnosed, our patient was treated with intravenous methylprednisolone sodium succinate, which was then switched to oral prednisone, hydroxychloroquine, methotrexate, and intravenous immunoglobulin. She was also prescribed calcium and vitamin D supplements, to improve overall bone density, and sunscreen that blocks UV-A and UV-B, to minimize the skin rash.

Early treatment with intravenous methylprednisolone and the early introduction of methotrexate and/or intravenous immunoglobulin have been used to achieve rapid control of the disease, to minimize treatment-related toxicity, and to reduce serious complications (like calcinosis).[4] Additional therapeutic agents include cyclosporin A, mycophenolate mofetil, systemic tacrolimus, rituximab, and cyclophosphamide. Topical steroids and immunomodulators, photoprotective agents, calcium, vitamin D supplements, and physiotherapy are all important adjunctive therapies to improve the outcome. The treatment is usually continued until improvements in clinical and laboratory results are achieved, and then the dosages are slowly reduced over the course of 2 to 3 years.

The prognosis for patients with JDM has improved in recent years. The mortality rate is 3.1%, and fewer than 10% of the children had significant disability.[5] More than half of these patients had chronic disease with increased frequency of calcinosis and lipodystrophy.[5]

Although our patient represents a classic case of JDM, the diagnosis was delayed because her skin finding was mistaken for eczema or an allergic skin reaction before she presented to our clinic. Practitioners should consider JDM in the differential diagnosis when treating patients with skin findings similar to eczema, especially when the rash does not respond to topical steroids or immunomodulators. Early diagnosis and treatment are very important for achieving rapid control of the disease and for providing a better long-term outcome for the patient.

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