



DERMATOMYOSITIS PRESENTING AS QUADRI-PARESIS A CASE REPORT VAASANTHI

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Abstract : Dermatomyositis, an idiopathic inflammatory myopathy, is one of the potentially treat-able causes of skeletal muscle weakness. The hallmark of this condition is its characteristic skin involvement which differentiates this from other inflammatory myopathy. Here we report a case of dermatomyositis who presented with flaccid quadri-paresis and bulbar involvement without classical dermatological signs.

Keyword: Dermatomyositis, inflammatory myopathy, autoimmune, perifascicular atrophy, heliotrope rash

CASE REPORT

31yr old female was admitted in our hospital with complaints of low grade fever for 2 days , facial puffiness, diffuse swelling of right side of neck & right upper limb. After a period of 10 days she developed difficulty in swallowing (both for solids and liquids), nasal regurgitation and slurring of speech and was put on ryles tube feeding. In a span of six weeks time she initially developed weakness of neck muscles, followed by upper limb and lower limb weakness. Her bladder and bowel habits were normal .No history of sensory involvement.

On general examination she had pallor, pedal edema , facial puffiness , macular rash over the neck. Her vitals were stable. Central nervous system examination revealed normal higher functions, nasal twang in speech , uvula in midline, diminished palatal movements, absent gag reflex. Spinomotor system: Mild edema of arms and thighs seen. Hypotonia in all 4 limbs. Both proximal and distal weakness was present along with neck muscle weakness. Deep tendon reflexes absent. Plantar flexor. Other system examination was normal.

Her investigations showed dimorphic anemia in peripheral smear. Chest xray , echocardiogram, renal and liver function test were normal. No abnormal findings in CT brain and CSF analysis. USG & Doppler of neck - moderate degree of interstitial edema present in the right side of neck. Venous flow normal. ANA was negative and HIV ELISA non reactive.

Serum CPK 1200 IU/L, EMG suggestive of myogenic pattern, MRI cervical spine showed swollen paraspinal muscles with increased intensity in favour of inflammatory myopathy.

Muscle biopsy from vastus lateralis revealed perifascicular atrophy with lymphocyte infiltration around blood vessels consistent with inflammatory myopathy probably dermatomyositis. fig 1



MRI cervical spine cross section

fig 2



MRI cervical spine showing increased signal intensities in paraspinal muscles

Patient was started with iv methylprednisolone and then maintained on methotrexate and oral prednisolone. She had a dramatic response to steroids , her muscle power improved and CPK became normal.

DISCUSSION:

Inflammatory myopathies are classified into three major groups: polymyositis, dermatomyositis, and inclusion body myositis. Dermatomyositis usually presents with skin and muscle symptoms. The predominant symptoms are muscle weakness and low muscle endurance. The weakness is most pronounced in proximal muscle groups—typically in the neck, pelvic, thigh, and shoulder muscles—with a symmetric distribution⁽³⁾. The weakness can be mild, moderate, or severe enough to lead to quadriplegia as in our case. Loss of tendon reflexes can be due to severe involvement of the muscles or due to peripheral nerve involvement⁽⁴⁾. Problems with swallowing and nutrition may occur as a result of impaired contractility of the throat muscles⁽⁵⁾, possibly leading to aspiration pneumonia. In rare cases, diaphragm or thoracic muscles might get involved. Though our patient had presented with a macular rash over the neck, the pathognomonic skin manifestations are Gottron's papules⁽⁶⁾ (slightly elevated violaceous, papules located over the dorsal side of the metacarpal or interphalangeal joints) and the heliotrope rash (periocular red or violaceous erythema of eyelids, often with edema). Other signs are erythematous rash over the neck (V sign)⁽⁷⁾, shoulders and back (shawl sign), hip (holster sign), periungual erythema, nail-fold telangiectasias, and cuticular overgrowth can also occur. This patient had presented with fever and limb edema before the onset of weakness. Literature evidence suggests that generalised or limb edema may be a feature of dermatomyositis⁽⁸⁾. Other features that can be present are systemic symptoms like malaise, weight loss, arthralgia, and Raynaud's phenomenon especially when associated with a connective tissue disorder. Cardiac and pulmonary involvement can also occur. Dermatomyositis specifically appears to have increased incidence of malignant conditions. The most common tumors associated are ovarian cancer, breast cancer, melanoma, colon cancer, and non-Hodgkin lymphoma. The presence of autoantibodies and the association with many autoimmune and connective tissue disorders favours an autoimmune etiology for inflammatory myopathy. In DM the endomysial inflammation is predominantly perivascular. The muscle fibers undergo necrosis, degeneration, and phagocytosis, due to microinfarcts within the muscle which results in perifascicular atrophy. The presence of perifascicular atrophy is diagnostic of DM, even in the absence of inflammation.

Bohan and Peter proposed a classification criteria for Polymyositis and Dermatomyositis which includes, Symmetric proximal muscle weakness, Increase in serum muscle enzymes, such as creatine kinase, Abnormal electromyographic findings, Abnormal muscle biopsy findings, Skin rashes. This condition responds to steroids and immunosuppressive drugs.

CONCLUSION

When it presents in its classical form, dermatomyositis is easy to diagnose, with typical rash over the knuckles and eyelids, along with a predominantly proximal upper and lower limb muscle weakness. However, in the absence of such characteristic manifestations, high index of clinical suspicion is required for early diagnosis and management. They should also be screened for associated connective tissue disease and malignancy.

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