AN INTERESTING CASE OF DERMATOMYOSITIS

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ABSTRACT

BACKGROUND

Dermatomyositis is a rare disease affecting the muscles along with the typical skin manifestations. It may be associated with visceral involvement and malignancy.

Case Presentation - We report the case of a 60-year-old man who had fever, rashes and weakness. Clinical diagnosis of primary muscle disease? Dermatomyositis was done; further confirmed by muscle biopsy.

CONCLUSION

The above case is presented for its rare incidence, atypically normal CPK enzyme levels and a good response to steroids.

KEYWORDS


BACKGROUND

Dermatomyositis is an uncommon systemic Rheumatic disorder characterised by inflammatory and degenerative changes in muscles and in the skin. Most specific signs are Gottron's papules over the knuckles and periorbital heliotrope rash, symmetric proximal limb girdle muscle weakness, tenderness and later atrophy. Complications include visceral involvement and cancer. Diagnosis is by clinical findings, increased muscle enzymes, electromyography and muscle biopsy.

CASE REPORT

A 60-year-old male presented with h/o high grade fever associated with chills 8 months back following which he developed pain in B/L thighs, buttocks and weakness of proximal muscles of the lower limbs. About 2 weeks later, he developed weakness of the proximal muscles of upper limbs. He then developed rashes on scalp, face, neck associated with swelling around the eyes. H/o dysphagia and nasal regurgitation present. He is a known case of hypothyroidism, on treatment. Clinically, primary muscle disease is suspected (Myopathy, myositis). On examination, patient had rashes. Vitals were normal. On nervous system examination, higher mental functions and cranial nerve examination were normal. Motor system examination revealed bulk and tone normal. Power of B/L upper limbs 4/5 around the shoulder and lower limbs 3/5 around the hip B/L.

DTRs were well preserved, B/L plantars were flexor. Sensory system and other systemic examination was unremarkable.
Clinical Diagnosis of Myositis? Dermatomyositis was made.

- Routine investigations were unremarkable.
- ESR – 62 mm at the end of 1 hour.
- Gastroduodenoscopy - Grade 1 Oesophageal varices.
- Creatinine phosphokinase was 33 IU/L
- Serum LDH - 52 IU/L.
- USG abdomen - small echogenic focus along the wall of gall bladder.
- EMG of right biceps and quadriceps - normal insertional activity with early complete recruitment.
- MUPs were of small amplitude with polyphasic potentials, s/o myopathic process.
- Muscle biopsy showed dermatomyositis (left biceps).
- Dermatomyositis was diagnosed and workup was done to rule out malignancy.

The above case is presented for its rare incidence, atypically normal CPK enzyme level and good response to steroids. Treatment with steroids was started, improvement in muscle power and skin changes were seen within 2 weeks of initiation of therapy.

DISCUSSION

Dermatomyositis is a distinctive entity identified by a characteristic rash accompanying, or more often preceding, proximal muscle weakness. The rash may consist of a blue-purple discoloration on the upper eyelids with oedema (Heliotrope). The erythematosus rash can also occur on other body surfaces, neck and anterior chest (often in a V sign), or back and shoulders (shawl sign). A definitive diagnosis is based on, Bohan and Peter classification, requiring a characteristic rash; plus three of the following:

1. Symmetrical, proximal muscle weakness.
2. Muscle biopsy abnormalities: Per fascicular atrophy.
4. EMG abnormalities.
5. Heliotrope rash or Gottron’s papules.

As with our case, the patient presented with pathognomonic cutaneous manifestations of heliotrope rash and poikiloderma along with proximal muscle weakness. The muscle biopsy abnormalities confirmed the diagnosis of DM.

The unique features drawn from this case are the dramatic response to the treatment and normal CPK levels.

Scan of the literature shows that the overall amelioration of the proximal muscle weakness and the rash with heliotrope takes minimum of three to six months following treatment. Many of the studies have shown to take five to six months for the remission of muscular weakness. However, our case showed a dramatic improvement of muscle power with regression of skin lesions within two weeks of starting treatment.

CPK levels are shown to be increased in most of the cases of dermatomyositis with active disease. Also, the CPK levels are monitored for its reduction in response to the commencement of the treatment. But nearly 4% of cases are shown to be having normal CPK levels especially when associated with other connective tissue disorders or malignancies. Our case had normal CPK levels with no association of other connective tissue disorders.

Dermatomyositis is well known to be associated with malignancies. Malignancy is often detected within the first year of onset of dermatomyositis with incidence being higher in patients aged more than 40 years. Many studies have reported dermatomyositis in strong association with colorectal malignancy, gastric, pancreatic and lung malignancies, adenocarcinomas, ovarian and breast malignancies, nasopharyngeal and bladder carcinos as, lymphomas. In the presenting case, there was no association with malignancies.

CONCLUSION

Dermatomyositis is a relatively rare disease with the incidence of 9.63 per million population. Diagnosis is usually done with high clinical suspicion with typical skin changes and muscle weakness further confirmed by investigations. The above case is reported because of its rarity, normal CPK levels despite muscle involvement and prompt response to steroids.

REFERENCES