CASE REPORT

Subcutaneous oedema of upper limbs heralding an aggressive form of dermatomyositis

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SUMMARY

Subcutaneous oedema is a rare presenting clinical manifestation of dermatomyositis. In this case, we report a 44-year-old man presenting with bilateral upper extremity predominant swelling and weakness. The proximal muscle weakness, dysphagia and presence of Gottron's papules as well as poikiloderma like skin changes led to the clinical diagnosis of dermatomyositis. He received aggressive treatment with high-dose glucocorticoids and required intravenous immunoglobulin with improvement in his symptoms.

BACKGROUND

Dermatomyositis is a systemic disease which is a subset of the idiopathic inflammatory myopathies.¹ It usually presents with proximal muscle weakness along with a myriad of dermatological findings.² Oedema can be seen as a dermatological manifestation in the eyelids and in cases of juvenile dermatomyositis.² However, generalised and/or limb oedema as a presenting sign of dermatomyositis has been reported in only 28 adult cases so far. We report an aggressive form of dermatomyositis which was characterised by limb oedema limited to upper extremities only.

CASE PRESENTATION

A 44-year African-American man with a history of polysubstance use and hyperlipidaemia presented with a 1-month history of gradual worsening of swelling involving both arms (figures 1 and 2) and dysphagia. In addition, he endorsed a 3-month history of alternating periorbital swelling of both eyes, proximal muscle weakness that was more pronounced in the arms compared with his legs and 30 pounds unintentional weight loss. On presentation, he had a blood pressure of 118/69 mm Hg, heart rate of 103 beats per minute, temperature of 98°F/36.7°C, respiratory rate of 18 and room air oxygen saturation of 93%. Physical examination revealed 3+ non-pitting oedema in both arms, and hypopigmented macules coalescing over the eyes (figures 3 and 4) and extending to the temples and neck. The patient had no evidence of heliotrope eruption or shawl rash. Muscle strength examination revealed 2/5 strength proximally and 5/5 strength distally in both arms and 3/5 strength proximally and 5/5 strength distally in both legs; severe weakness of neck extensor muscles with the patient unable to support his head. The remainder of the examination was unremarkable.

INVESTIGATIONS

Laboratory examination was noteworthy for elevated creatine kinase levels of 16 800 (0–200 U/L), aldolase 59.2 (≤ 8 U/L), positive anti-nuclear antibody 1:640 ($< 1:40$), SSA (RO) antibody: 7.5 (< 1), elevated liver enzymes aspartate transaminase: 765 (0–40 IU/L), alanine transaminase: 215 (0–44 IU/L), low complements C3: 79 (86–184 mg/dL), C4: 17 (20–59 mg/dL), elevated inflammatory markers erythrocyte sedimentation rate: 45 (0–15 mm/hr) and C reactive protein: 1.23 (< 0.5 mg/dL). MRI of both arms showed diffuse hyperintensity representing muscle oedema (figures 5 and 6). Only three myositis-specific antibodies including Anti Jo-1, Anti-Mi-2 and anti-SRP were checked in our patient and found to be negative. Cancer screening including CT chest, abdomen-pelvis, upper endoscopy and nasopharyngolaryngoscopy did not suggest any malignancy. Other pertinent lab studies included negative anti-neutrophil cytoplasmic antibodies, HIV, urine drug screen and hepatitis panel. He was diagnosed with possible dermatomyositis. Diagnosis was based on findings of proximal muscle weakness, history of periorbital oedema, poikiloderma (hypopigmented rash), elevated creatine kinase levels in the absence of other possible causes and MRI of both arms consistent with diffuse muscle oedema.

DIFFERENTIAL DIAGNOSIS

Other causes of myositis besides dermatomyositis, deep venous thrombosis, hypothyroidism, renal failure, liver failure and heart failure.

TREATMENT

The patient was treated with pulse dose steroids 1 mg/kg for 3 days and later transitioned to 80 mg oral prednisone daily. There was no change noticed in dysphagia, oedema and strength in both arms after high-dose prednisone use. The decision was made to administer intravenous immunoglobulin (Ig), 400 mg/kg daily for 5 days (total dose of 2 g/kg) and percutaneous endoscopic gastrostomy tube was placed for ongoing dysphagia. He was discharged to a rehabilitation centre on 40 mg of oral prednisone.

OUTCOME AND FOLLOW-UP

The patient was seen at 1 week and 4 weeks interval after hospital discharge. At the 1-week appointment, the muscle strength slightly improved with 3/5 in his upper extremities proximally and 4/5

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Figure 1 Patient at presentation with bilateral upper extremity swelling only.

strength proximally in his lower extremities but still unable to hold his neck erect. He still had 2+ non-pitting oedema in both arms and was still dependent on tube feeds due to ongoing dysphagia. He was continued on 40 mg prednisone with 10 mg taper over next 2 weeks and low-dose azathioprine 50 mg daily was started.

At 4 weeks, he was seen in the hospital after he was admitted after an aspiration event at the rehabilitation centre. The examination this time revealed that muscle strength improved

Figure 2 Note the size difference of arms as compared with the legs depicting swelling only limited to arms.

Figure 3 Hypopigmented rash over the right side of the eye and neck.

significantly at this encounter 4/5 in his upper extremities proximally and 4/5 strength proximally in his lower extremities, neck muscles strength 5/5 and oedema in the hands resolved (figure 7). He was maintained at 10 mg prednisone, azathioprine was held (due to infection concerns) and the creatine kinase level was noted to be normal 76 (0–200 U/L).

The patient is a piano player and after 5 weeks of treatment, swelling of the arms was completely gone and he regained his strength back to play the piano!

DISCUSSION

Subcutaneous oedema is a rare presenting manifestation of dermatomyositis and our case will be the 29th case to date. Patients generally present within days–weeks with weakness along with dysphagia and elevated creatine kinase levels. Prior cases have had oedema as the only dermatological manifestation of dermatomyositis and may be a sign of a severe form of the disease that requires early and aggressive therapy.² These cases of oedema are attributed to dermatomyositis once other underlying causes of oedema such as liver, renal, cardiac disease, hypothyroidism and deep venous thrombosis are ruled out.^{2,3} The reason why limb oedema is limited to upper limbs is unclear. The mechanism by which dermatomyositis causes oedema is not well understood either; however, it is suggested that muscle inflammation causes the capillaries to leak, which in turn causes oedema within the muscles and by extension throughout the body.³ Another theory is that deposition of activated complement within muscle capillaries causes microinfarcts and perivascular inflammation, leading to increased permeability and causing oedema.⁴ Dermatomyositis-associated antibody antinuclear matrix protein-2

Figure 4 Hypopigmented rash over the left eye extending to the temple.

autoantibodies (anti-NMP-2 Ab) have been described to be associated with subcutaneous oedema, more severe form of muscle disease and increased malignancy risk.⁵ Anti-NMP-2 Ab was not checked in our case because of lack of awareness of this antibody relationship with our case at the time of management. Myositis specific 11 antibodies panel that also includes anti-NMP-2 Ab is

Figure 5 MRI image T2-weighted sagittal view showing diffuse hyperintensity throughout all the muscles consistent with the diagnosis of myositis.

Figure 6 MRI image STIR coronal view showing diffuse hyperintensity throughout all the muscles consistent with the diagnosis of myositis.

Figure 7 Resolution of the upper limb oedema after 4 weeks of treatment. Compare the size of the arms before and after treatment.

commercially available and usually a send-out test when ordered in the inpatient settings.

In most patients with dermatomyositis presenting with oedema, many recovered after being treated with either one of three regimens: oral prednisone, prednisone with either methotrexate or azathioprine, or prednisone with intravenous Ig.^{2,6} It has also been shown that intravenous Ig is effective in cases with bulbar involvement.⁷ In our case, the patient presented with upper extremity weakness and oedema for 1 month along with dysphagia. Our patient was worked up for other signs of oedema and all studies were negative. The absence of other diseases that can cause peripheral oedema along with the improvement in patient's symptoms with glucocorticoids with intravenous Ig suggested that the oedema was due to dermatomyositis. Our patient's dysphagia also improved with intravenous Ig, something that had worked in other cases.⁷

Learning points

- ▶ Subcutaneous oedema of limbs can be a rare presenting sign of severe form of an adult dermatomyositis.
- ▶ This form of dermatomyositis appears to be more aggressive in nature and potentially have an increased malignancy risk.
- ▶ Patients with this form of dermatomyositis usually require high-dose prednisone 1 mg/kg oral or intravenous, prednisone with either methotrexate or azathioprine, or prednisone with intravenous immunoglobulin.
- ▶ Intravenous immunoglobulin is effective in cases of dermatomyositis with dysphagia.

Contributors MS: performed the literature search and discussion. RP: performed clinical case presentation. HK: was the attending physician who acted in the co-management of this clinical case and was also reviewer of the case report. DF: provided his expertise in reviewing as well as preparation of the draft.

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