A Possible Case of Systemic Lupus Erythematosus Presenting with Generalised Oedema

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Abstract: Systemic lupus erythematosus (SLE) is an autoimmune disease of unknown aetiology affecting various systems within the body. We report the case of a patient with generalised subcutaneous oedema as the only presenting feature, which led to the possible diagnosis of SLE without a specific cause. The patient presented to the Sultan Qaboos University Hospital in Muscat, Oman, in April 2013. The oedema had been present for two years before admission. Other potential causes of oedema in patients with SLE were excluded, including SLE of renal origin and SLE due to protein-losing enteropathy or drugs. This was confirmed by the patient’s normal serum albumin level and negative proteinuria. Laboratory investigations showed high levels of positive antinuclear antibodies (1:1,640), positive anti-double stranded DNA, high levels of anti-beta2-glycoprotein 1 and immunoglobulin M, and low levels of both complement components 3 and 4. The oedema improved immediately in response to steroids and immunosuppressive medications. Physicians should be aware that generalised subcutaneous oedema can be the only manifestation of SLE.

Keywords: Edema; Systemic Lupus Erythematosus; Case Report; Oman.

Case Report

A 21-year-old unmarried female university student with no previous medical problems presented to the Sultan Qaboos University Hospital in Muscat, Oman, in April 2013 with symptoms of generalised swelling of the body which had been present for two years. The swelling was located mainly in the face, abdomen and lower limbs and was gradually increasing over time. The swelling was at its worst in the morning, to the extent that sometimes she was unable to open her eyes fully for 30 minutes after waking. Over the two-year period, the patient noted that her weight had increased by 17 kg; she had begun regular exercise, but had not succeeded in losing any weight. The patient denied experiencing any of the following symptoms: joint pain, chest pain, shortness of breath, palpitations, dizziness, skin rashes, oral ulcers, hair loss, changes in appetite or menstrual problems. There was no history of allergies or any significant family history. The patient was not currently taking any medication.

A physical examination revealed puffiness of the face and pitting pedal oedema extending up to the toes. On further questioning, the patient reported experiencing swelling of the face and pitting pedal oedema for two years prior to admission, which had been considered a normal part of her body due to her ethnic background. She denied experiencing any of the following symptoms: joint pain, chest pain, shortness of breath, palpitations, dizziness, skin rashes, oral ulcers, hair loss, changes in appetite or menstrual problems.

Laboratory investigations showed high levels of positive antinuclear antibodies (1:1,640), positive anti-double stranded DNA, high levels of anti-beta2-glycoprotein 1 and immunoglobulin M, and low levels of both complement components 3 and 4. The oedema improved immediately in response to steroids and immunosuppressive medications. Physicians should be aware that generalised subcutaneous oedema can be the only manifestation of SLE.
There are reports of remitting asymmetrical pitting oedema in this patient, as patients with protein-losing enteropathy (PLE) was unlikely to be the cause of the generalised oedema. Moreover, bilateral pleural and pericardial effusions from Sjögren's syndrome, as a cause of oedema in this patient was excluded by the absence of urinary protein. Moreover, urinary protein and proteinuria were excluded in this patient. Additionally, the patient’s autoimmune profile was strongly positive for blood anti-cardiolipin antibody (immunoglobulin G) and anti-cardiolipin antibody (immunoglobulin M), and the patient had positive ANA and anti-n-DNA antibodies recorded. The current patient was SLE, due to the high levels of anti-n-DNA antibodies. The investigation results suggested a diagnosis of SLE. However, her clinical condition fulfilled neither the diagnostic criteria of SLE (according to the revised guidelines of the American College of Rheumatology) nor those of mixed connective tissue disease. The diagnosis was found to be SLE based on immunological findings. Since her autoimmune profile was strongly indicative of SLE, it was thought likely that this was a possible case of SLE, potentially progressing to definite SLE in the future. The response of the oedema to immunosuppressive therapy also supported this diagnosis.

More common causes of subcutaneous oedema (such as heart failure, liver disease, malnutrition, renal disease or drugs) were excluded in this patient by a careful history-taking as well as clinical and other relevant investigations. Oedema, especially the localised form, has been reported in the literature as a rare presenting symptom of SLE. There are reports of periorbital oedema, lower limb pitting oedema, facial oedema, remitting asymmetrical pitting oedema and angioedema, all as initial presentations of SLE. There are several cases of SLE presenting with generalised oedema due to either protein-losing enteropathy (PLE) or polyserositis in the form of massive bilateral pleural and pericardial effusions. Nephrotic syndrome as a cause of oedema in this patient was excluded by the absence of urinary protein. Moreover, PLE was unlikely to be the cause of the generalised subcutaneous oedema in this patient.
PLE usually have low serum protein and albumin measurements.\textsuperscript{10,12} As a result, tests to detect PLE, such as technetium\textsuperscript{99m} albumin scintigraphy or a 24-hour stool alpha-1-antitrypsin clearance test, were not justified in this case.\textsuperscript{7,8}

The underlying cause of generalised oedema in SLE patients without systemic manifestations, such as renal disease, is not yet clear. Günaydin \textit{et al}. postulated that the localised oedema observed in his reported case was most likely due to vasculitis, which had led to an obstruction of the lymphatic vessels.\textsuperscript{6} The aetiology of localised periorbital oedema in patients with SLE flares is also not apparent. A case series reported by Gómez-Puerta \textit{et al}. found that while some cases were related to nephrosis, there was no evidence in others.\textsuperscript{14} Pittau \textit{et al}. believed that oedema may be due to a transient impairment in lymphatic drainage or pre-existing increased capillary permeability, as demonstrated in patients with connective tissue disorders.\textsuperscript{8} Marks \textit{et al}. proposed that there was an increase in vascular permeability in patients with connective tissue diseases.\textsuperscript{15} In yet another patient with periorbital oedema, increased dermal mucin deposits were observed during a biopsy.\textsuperscript{16} Angioedema due to C1-inhibitor deficiency has also been described in the literature.\textsuperscript{17}

The findings of this case report are limited by certain factors. Photographs of the patient were not taken before the initiation of treatment and a skin biopsy was not sent to a pathologist to determine the exact pathophysiological mechanisms of the patient’s symptoms.

Conclusion
The first possible case of SLE presenting with generalised subcutaneous oedema without a definite cause is described. The cause of oedema in this patient could not be explained by other reported causes of generalised oedema associated with SLE, such as PLE, nephrotic syndrome or polyserositis. The presence of generalised oedema is a rare cutaneous manifestation of SLE and can be the initial and sole manifestation of this disease.

References