Dropped head: an unusual presentation of dermatomyositis.

Gleb Slobodin, Michael Rozenbaum, Boaz Weller, Nina Boulman and Itzhak Rosner

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Subclinical Atherosclerosis in Systemic Lupus Erythematosus

To the Editor:

We read with interest the editorial by Ahmed and Bruce and the article by Wolak et al. We recently used carotid artery ultrasonography in premenopausal female patients with systemic lupus erythematosus (SLE) and in age-matched controls, to detect carotid plaques, to evaluate risk factors associated with atherosclerosis, and to identify clinical and laboratory variables related to the presence of carotid plaques in these women.

Our study included 26 SLE patients and 20 female controls who underwent carotid ultrasound. All the SLE patients met American College of Rheumatology criteria for the classification of SLE. Carotid intima media wall thickness (IMT) was measured in both left and right common carotid arteries using an HDI 1500 ( ATL, Bothell, WA, USA) equipped with a 12.5 MHz vascular transducer. The main clinical features of the lupus patients at any time during the disease were: pancytopenia, serositis, polyarthritis, hemolytic anemia, thrombocytopenia/leukopenia, renal failure, nephrotic syndrome, cerebrovascular disease, cognitive dysfunction, and photosensitivity.

At the time of ultrasonography study, lupus patients were receiving oral prednisone, azathioprine, nonsteroidal antiinflammatory drugs, cytotoxic drugs, and hydroxychloroquine. Demographic and atherosclerotic risk factors of patients with SLE and controls are shown in Table 1. Compared with controls, SLE patients had greater carotid IMT, total cholesterol, triglycerides, and hypertension. Age, body mass index (BMI), LDL cholesterol, HDL, and C-reactive protein (CRP) of patients and controls were not statistically different. Seven (26.9%) of 26 patients showed carotid plaques, compared with only one (5%) of 20 controls. One patient from each group had past or present smoking history.

Comparison of patients with and without carotid plaques (Table 2) showed a significant difference only in age (41.8 vs 30.6 years; t test, p = 0.006). Five of 7 patients with carotid plaques had moderate levels (20–80 U) of IgG cardiolipin antibodies (Relisaa Cardiolipin IgG and IgM, Immuno Concepts, Sacramento, CA, USA) and 2 had low levels (5–20 U). Of the 7 patients with carotid plaques none had received cytotoxic drugs, while 7 of 19 patients without plaques had received cytotoxic agents (Fisher's exact test, p = 0.000). Variables significantly associated with carotid plaques by logistic regression analysis included age (OR 5.15, 95% CI 0.36–72.7, p = 0.001) and presence of anti-cardiolipin antibodies (OR 1.38, 95% CI 0.82–2.3, p = 0.008).

In our study, subclinical atherosclerosis in premenopausal SLE patients was not related to traditional cardiovascular risk factors. Clinical or aorto-logic measures of disease activity or serum levels of CRP were not significantly different, although all measures were obtained at the time of ultrasonography. As reported by Romano, et al. and Assman, et al., we also found that long-term therapy with corticosteroids was not associated with a significantly increased risk for carotid atherosclerosis. Plaques were more common among patients who had not received aggressive therapy with cytotoxic drugs than in those who had. Finally, a logistic regression model identified age and antiphospholipid antibodies as risk factors for atheroma.

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REFERENCES

Table 1. Demographic and atherosclerotic risk factors of patients with SLE and controls. Values are mean ± standard deviation (SD).

<table>
<thead>
<tr>
<th></th>
<th>SLE, n = 26</th>
<th>Controls, n = 20</th>
<th>p</th>
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<tbody>
<tr>
<td>Age, yrs</td>
<td>33.6 ± 9.7</td>
<td>29.2 ± 6.0</td>
<td>0.08</td>
</tr>
<tr>
<td>Body mass index, kg/m² body surface</td>
<td>25.6 ± 6.0</td>
<td>25.0 ± 4.1</td>
<td>0.70</td>
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<tr>
<td>LDL cholesterol, mg/dl</td>
<td>110.3 ± 35</td>
<td>95.7 ± 20.4</td>
<td>0.10</td>
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<tr>
<td>Total cholesterol, mg/dl</td>
<td>183.5 ± 40</td>
<td>159.6 ± 25</td>
<td>0.02</td>
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<tr>
<td>HDL, mg/dl</td>
<td>43.8 ± 13</td>
<td>49.4 ± 10.4</td>
<td>0.11</td>
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<tr>
<td>Triglycerides, mg/dl</td>
<td>145.3 ± 68</td>
<td>74.5 ± 40</td>
<td>0.001</td>
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<tr>
<td>Systolic blood pressure, mm Hg</td>
<td>122.7 ± 19</td>
<td>108.7 ± 8</td>
<td>0.004</td>
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<tr>
<td>Diastolic blood pressure, mm Hg</td>
<td>79.3 ± 11</td>
<td>69.5 ± 8</td>
<td>0.001</td>
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<tr>
<td>Intima media thickness, mm</td>
<td>0.62 ± 0.11</td>
<td>0.48 ± 0.05</td>
<td>0.001</td>
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<tr>
<td>Left common carotid artery, mm</td>
<td>0.55 ± 0.08</td>
<td>0.46 ± 0.05</td>
<td>0.001</td>
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<tr>
<td>Right common carotid artery, mm</td>
<td>6.9 ± 3.7</td>
<td>6.0 ± 0</td>
<td>0.25</td>
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<tr>
<td>Carotid plaques (%)</td>
<td>7 (20.9%)</td>
<td>1 (5%)</td>
<td></td>
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<tr>
<td></td>
<td>With Plaques, n = 7</td>
<td>No Plaque, n = 19</td>
<td>p</td>
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<tr>
<td>Age, yrs</td>
<td>41.8 ± 4.4</td>
<td>30.6 ± 9.4</td>
<td>0.006</td>
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<tr>
<td>Body mass index, kg/m² body surface</td>
<td>25.4 ± 1.1</td>
<td>25.8 ± 8</td>
<td>0.89</td>
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<td>Total cholesterol, mg%</td>
<td>167.9 ± 23.9</td>
<td>189.3 ± 43.5</td>
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<tr>
<td>LDL cholesterol, mg%</td>
<td>99.3 ± 17.5</td>
<td>114.5 ± 39</td>
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<tr>
<td>Anticardiolipin (GPL)</td>
<td>31.4 ± 22</td>
<td>21.2 ± 24</td>
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<tr>
<td>Anticardiolipin (MPL)</td>
<td>5.0 ± 0.0</td>
<td>11.4 ± 14.2</td>
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<td>Systolic blood pressure, mm Hg</td>
<td>122.3 ± 19.2</td>
<td>123 ± 20</td>
<td>0.94</td>
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<td>Diastolic blood pressure, mm Hg</td>
<td>79 ± 13</td>
<td>79 ± 10.4</td>
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<td>Anti-DNA*, IU/ml</td>
<td>16.7 ± 17.2</td>
<td>31.5 ± 19.6</td>
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<td>SLICC</td>
<td>0.14 ± 0.38</td>
<td>0.95 ± 1.7</td>
<td>0.22</td>
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<td>Mex-SLEDAI</td>
<td>2.4 ± 2.3</td>
<td>2.8 ± 2.3</td>
<td>0.72</td>
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<td>SLE duration, yrs</td>
<td>10.1 ± 4.5</td>
<td>6.4 ± 6.0</td>
<td>0.13</td>
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<tr>
<td>Prednisone, mg</td>
<td>8.6 ± 11.9</td>
<td>9.3 ± 7.3</td>
<td>0.84</td>
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<tr>
<td>C-reactive protein, mg/l</td>
<td>8.6 ± 6.8</td>
<td>6.3 ± 1.4</td>
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</table>


Drs. Ahmad and Bruce reply

To the Editor:

We thank Drs. Mercado and Avendaño for their interest in our recent editorial and for sharing their own data relevant to this question. Their comparison between systemic lupus erythematosus (SLE) and controls is typical of many other studies in that they find an excess of metabolic risk factors in their SLE population as well as a higher prevalence of markers of early atherosclerosis.

In agreement with our opinion, they found that increasing age was the most significant factor associated with plaque development in SLE patients. Their data also suggest a contribution to atherosclerotic risk of the presence of antibodies to cardiolipin as well as a lack of previous exposure to cytotoxic therapy. These latter 2 observations are of particular interest. Controversy continues as to the importance of anticardiolipin antibodies to atherogenesis in SLE, but several studies suggest that certain subtypes of anticardiolipin antibody may be of particular relevance. This area needs further attention. Equally, the role of aggressive therapy for SLE is of intense current interest and it may be that such aggressive therapy early in the disease "resets" the vasculature in a way that makes it less susceptible to the development of atherosclerosis in the long term.

Clearly, larger studies are required to delineate further the contribution of lupus associated factors to atherosclerosis. Our colleagues' preliminary data add some intriguing observations to our body of knowledge.

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REFERENCES


Dr. Abu-Shakra replies

To the Editor:

It is no longer a matter of dispute that patients with systemic lupus erythematosus (SLE) have an increased risk of developing clinical and asymptomatic cardiovascular disease, particularly before the age of 50. The results of the work of Drs. Mercado and Avendaño support our published data. However, we identify differences between the studies.

Our controlled study showed that patients with SLE have a 3.17-fold increased risk of atherosclerotic plaques in the carotid and femoral arteries compared with controls. Drs. Mercado and Avendaño report that SLE patients had a higher rate of carotid plaques and greater carotid intima media thickness (IMT) compared to controls. Our data and data of a large study from New York have not shown a difference between SLE cases and controls in the measurements of IMT.

This difference is related mainly to sample size and selection bias. In Mercado and Avendaño’s study only 26 SLE patients were included and the number of controls was 20. This discrepancy suggests that there were SLE patients without matched controls. Mean total cholesterol, triglyceride level, and mean systolic and diastolic blood pressure were significantly higher in SLE cases versus controls, indicating that cases and controls were not tightly matched by conventional atherosclerosis risk factors. In addition, there was a trend suggesting controls were younger than cases by 4 years. All of these variables most likely contributed to the higher IMT measurements observed in the SLE patients.

We have shown in a multivariate model that the development of plaques is associated with age, particularly in those older than 50 years (odds ratio 2.66, p < 0.001). The authors report that plaques were significantly associated with age and presence of anticardiolipin antibodies. This should be interpreted with caution, since 1 is included in the range of the 95% confidence intervals of the odds ratios. Again only 7 SLE patients had...
Temporal Arteritis Associated with Systemic Necrotizing Vasculitis

To the Editor:

I read with interest the report of 7 cases of temporal arteritis in patients with systemic vasculitis without classic giant cell arteritis. Non-giant cell arteritis, such as necrotizing vasculitis, polyarteritis and “hypersensitivity” angitis, have been reported to involve the temporal arteries much earlier than the reports quoted by Hamidou et al. The prognosis of those patients with non-giant cell arteritis may be less favorable, probably due to the less beneficial effect of single-corticosteroid therapy on the course of polyarteritis. Clinicians and histopathologists should be aware of the possibility of non-classic giant cell arteritis in elderly patients presenting with temporal arteritis. A temporal artery biopsy should be performed in patients with temporal arteritis, as patients with systemic necrotizing vasculitis may require cytotoxic drugs in addition to corticosteroids.

I reported the case of a female patient who, at the age of 50 years, developed biopsy-proven classic polyarteritis nodosa, from which she made a full recovery, and was taken off corticosteroids 22 months after onset. Sixteen years later, she developed classic giant cell arteritis with typical histological findings. This patient had characteristic clinical and histological findings of both diseases, but at a 16-year interval. This may be a chance finding.

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REFERENCES


Dr. Hamidou replies

To the Editor:

We thank Dr. Jawad for his remarks on non-giant cell temporal arteritis. The overall prognosis of these patients may be effectively less favorable if other sites are involved, and clinicians must be aware of renal, cardiac, digestive, or central nervous system involvement, defining requirements of the “five factors score” and requiring cytotoxic drugs. Curiously, in the literature and in our experience, visual disturbances are rare in these settings and blindness exceptional. Histopathologically, fibrinoid necrosis of temporal artery is unusual in classical giant cell arteritis, and it could be a suggestive element of systemic necrotizing vasculitis. Several descriptions of association of giant cell temporal arteritis and necrotizing vasculitis have been reported, without any proved pathogenic relationship.

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REFERENCES


Silicone Breast Implants

To the Editor:

The 2 letters in the May issue of The Journal about breast implants are unscientific and silly. They point up the variation that can occur even in a series, and refer to subjective reports that have no meaning. Chronic fatigue syndrome and fibromyalgia cannot be verified and women who have cosmetic breast implants differ from those who don’t, and those who have implants after mastectomy have other things to worry about and cannot be compared to women who have not had cancer. The article under discussion and the letters in response serve to confuse and add nothing to our knowledge. Let’s get back to science-based medicine.

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REFERENCES


Dr. Vermeulen replies
To the Editor:
We thank Dr. Ehrlich for his contribution to the scientific discussion.

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Dropped Head: An Unusual Presentation of Dermatomyositis
To the Editor:
Muscle weakness is a cardinal feature of idiopathic inflammatory myopathies that triggers further investigations and proper diagnosis by high serum levels of muscle enzymes, characteristic electromyography (EMG), and muscle biopsy. Most patients with bilateral, symmetric, progressive, proximal weakness leading to difficulty getting up out of a chair or combing their hair are usually diagnosed within weeks or months from onset of complaints, while those presenting with atypical weakness may go undiagnosed, and subsequently untreated, for years.

We describe a case of dermatomyositis with prominent weakness of axial musculature and resultant dropped head syndrome. While neck flexor involvement with difficulty in raising one’s head from a pillow is a known manifestation of inflammatory myopathy, a weakness of neck extensors with dropped head has not been previously reported, to our knowledge, as a presenting feature of dermatomyositis.

A 66-year-old Caucasian woman first presented in March, 2003, with a 4-month history of difficulty in maintaining her head erect. Her muscle strength was estimated as 2/5 for neck extensors and 4/5 for deltoid muscles, with other muscle groups normal. EMG revealed C6 radiculopathy with no myopathic changes. Computer tomography (CT) and later magnetic resonance imaging (MRI) of the C-spine showed degenerative changes of the C3-C7 segments with minimal spinal stenosis, which was not felt to explain the patient’s complaints. Final diagnosis was deferred.

In September 2003 the patient was reevaluated because of worsening weakness of the neck with complete inability to hold her head up and proximal weakness of both upper and lower extremities. She had a waddling gait with dropped head and declined her torso in a sway back posture to allow her to see where she was walking. The review of systems revealed...
mild dysphagia, Raynaud's phenomenon, and weight loss of about 20 kg since onset of symptoms 9 months earlier. Her medications were amitriptyline 25 mg, cimetidine 400 mg, and calcium carbonate 600 mg. Physical examination was remarkable for complete head drop, weakness of the neck extensors 0/5, left deltoid 3/5, right deltoid 4/5 and bilateral ilopsoas 3/5, violaceous rash over her forehead, upper back, chest, thighs, metacarpophalangeal and proximal interphalangeal joints, and lesions compatible with Gottron papules. She had periungual telangiectasia and taut skin on her distal fingers.

Routine laboratory studies, including complete blood count, serum creatinine, electrolytes, proteins, thyroid and liver function tests, were normal. CPK level was 419 U/I (normal range up to 100 U/I), lactate dehydrogenase was 344 U/I (normal range up to 240 U/I). C-reactive protein was 9.3 (normal up to 6) and erythrocyte sedimentation rate was 55 mm/h. Antinuclear antibody test was weakly positive in a nucleolar pattern, while extractable nuclear antigen (ENA) screen including anti-Jo-1 antibodies, as well as rheumatoid factor and antibodies to acetylcholine receptor were negative; serum complement levels C3 and C4 were normal. Extensive investigations to rule out underlying malignancy were negative. EMG of the right deltoid, splenius capitis, and semispinales muscles showed no spontaneous activity with fast recruitment of small, short, and polyphasic units. A biopsy of the left deltoid muscle was performed and revealed atrophic degenerative and necrotic fibers with foci of inflammatory infiltrate, composed of both T and B cells, compatible with dermatomyositis.

Dermatomyositis was diagnosed and the treatment with 1 mg/kg/day steroids and 15 mg of methotrexate (MTX) weekly was started. Two months later, monthly intravenous immunoglobulin (IVIG) was added to her treatment due to failure to improve. The patient reported gradual improvement in muscle strength since starting IVIG, which allowed tapering of her steroid dose. On reevaluation in June, 2004, she was able to hold her head straight up with no help and had returned to an active lifestyle. Her rash had disappeared and serum muscle enzyme levels were normal.

Restricted or isolated weakness of the extensors of the neck is a relatively rare condition, designated the dropped head syndrome, generally seen as a part of generalized neuromuscular disorder such as myasthenia gravis or amyotrophic lateral sclerosis. The dropped head is not widely described as a manifestation of rheumatic disease, despite the presence of inflammatory myopathies in the list of differential diagnoses. Reported cases of dropped head syndrome, characterized by the predominant weakness of neck extensors with restricted involvement of other proximal musculature, and biopsy varying from scattered interstitial inflammatory infiltrate to rich lymphocytic infiltration, may represent an early stage of polymyositis. These patients may have a good response to glucocorticosteroids and/or azathioprine with improvement of their muscle strength.

This case shows that the dropped head syndrome may also be an early and disabling symptom of dermatomyositis, still reversible with aggressive treatment. Awareness of such an unusual manifestation of inflammatory myopathy should advance diagnosis and antiinflammatory therapy and improve the overall prognosis of such patients.

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