Dermatomyositis as initial presentation of Hodgkin’s lymphoma: A case report and review of literature

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Introduction

The increased incident of malignancy is well established in patients with inflammatory myopathies and specifically in Dermatomyositis (DM). However, the association between DM and Hodgkin’s Disease (HD) is very unusual. We have recently had the opportunity of observing a patient who manifested the clinical features of DM and found to have HD after a detailed work up.

Case report

This 30 year old lady presented with history of fever for 2 months, which was initially low grade but later increased up to 104 F. Fever was intermittent and associated with night sweats. She had polyarthralgia involving bilateral symmetrical hand joints, wrists, knees and ankles but without swelling or early morning stiffness. She had characteristic skin lesions of DM including heliotrope eruption with violaceous erythema and mild edema of upper eyelids, erythematous to violaceous papules and plaques over both knuckles (Gottron’s papules), erythematous rash over upper chest (V sign) and upper back (shawl sign) and photosensitive papules over forearms and dorsum of hands.

On examination she had pallor, cervical and axillary lymphadenopathy and hepatosplenomegaly. Joint examination did not reveal active synovitis. Motor examination showed muscle power of 4/5 at hip flexion and extension. Rest of the limb and neck muscles were strong. Deep tendon reflexes were 2+ and bilateral plantars were flexor.

Investigation showed anemia with hemoglobin of 6.5 g/dl and peripheral blood film revealed normocytic normochromic red cells. Renal function test, liver function test and serum calcium were normal. Liver was 17.5 cm and spleen was 16.9 cm in size on ultrasonography.

Lactate Dehydrogenase (LDH) was 3880 U/L (reference values 240-480), Creatinine Kinase – Skeletal Muscle (CK-MM) was 344 U/L (reference values 10-165) and Electromyography (EMG) from right vastus lateralis and deltoid showed myopathic pattern. A muscle biopsy showed foci of perivascular lymphomononuclear inflammatory infiltrate with fiber regeneration and few of the fibers show muscle fiber splitting. Overall features were suggestive of myopathy pattern.

Skin biopsy from dorsum of hand lesion showed basal cell vacuolar degeneration with occasional apoptotic keratinocytes in epidermis and mild perivasculare chronic inflammatory cells, consistent with DM.

On the basis of characteristic skin rash distribution, elevated muscle enzymes (CK-MM, LDH), EMG, skin and muscle biopsy, a diagnosis of DM was made according to Bohan and Peter classification criteria1,2 DM associated connective tissue disease (rheumatoid arthritis, systemic lupus erythematosus, systemic sclerosis, Sjogren syndrome, mixed connective tissue disease) work up was negative. Anti-Jo1 antibody was negative and spirometry and computed tomography of chest did not show features of interstitial lung disease.

Work up for associated malignancy was carried out. Breasts and pelvis were normal on physical examination and on ultrasonography. In view of presence of lymphadenopathy and hepatosplenomegaly, a possibility of lymphoma was kept and a whole-body Positron Emission Tomography (PET) scan was done which showed generalized lymphadenopathy (bilateral cervical, bilateral axillary, bilateral hilar, peripancreatic, splenic, aortocaval, paraaortic, bilateral common iliac, internal and external iliac and bilateral inguinal lymph nodes) with a possibility of lymphomatous etiology. Left cervical lymph node fine needle aspiration cytology and right axillary lymph node biopsy showed reactive lymphoid hyperplasia. Bone marrow biopsy and splenic aspirate cytology were also suggestive of reactive changes. In view of a high possibility of lymphoma, a repeat lymph node biopsy from right cervical region was done which was consistent with HD, nodular sclerosis.

Ann Arbor staging was stage IV S B and ECOG (Eastern Cooperative Oncology Group) performance scale was 2. For HD ABVD (doxorubicin, bleomycin, vinblastin and dacarbazine) regimen was given and for dermatomyositis, she was started on oral prednisolone 1mg/kg and topical fluticasone (for skin lesions). Following these measures, her fever subsided, skin lesions improved and performance scale improved to ECOG 1 and LDH decreased to 1590 U/L on a subsequent outpatient follow up.

Discussion

DM is a systemic inflammatory myopathy characterized by...
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In all the cases, clinical features of DM appeared before or concomitant with the manifestation of HD, but never after. Temporal association does not imply a cause-effect relationship, however, the longer the time interval between the two diagnoses, the weaker becomes the correlate. Most solid cancers develop within one year of the onset of DM, although the risk for malignancy remains high for up to five years after a diagnosis of DM was established. [29]

The clinical features of DM are similar in the three groups of patients - those with no associated malignancy, those with HD, and those with other malignancies.

Regarding the extent and staging of the HD, eight patients (and index case) had stage III or IV and this proportion was higher than in a population with HD alone. Four patients had stage I-II and for four patients no stage was stated. It has been postulated that the advanced stage can be due to delay in the clinical diagnosis of the HD by steroid, started for the treatment of DM [21]. However the HD stage mentioned in the case reports is not necessarily is stage at initial diagnosis.

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Table 1. DM and HD: review of literature

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age/sex</th>
<th>Interval between the diagnoses (DM → HD)</th>
<th>Treatment of DM</th>
<th>Stage of HD</th>
<th>Treatment of HD</th>
<th>Outcome (duration of follow up)</th>
</tr>
</thead>
<tbody>
<tr>
<td>9</td>
<td>32/M</td>
<td>27 months</td>
<td>(ND)</td>
<td>(ND)</td>
<td>RT</td>
<td>Remission (ND)</td>
</tr>
<tr>
<td>10</td>
<td>59/M</td>
<td>2 months</td>
<td>CS</td>
<td>I/II</td>
<td>(ND)</td>
<td>Improved (3 months)</td>
</tr>
<tr>
<td>11</td>
<td>49/F</td>
<td>7 months</td>
<td>CS</td>
<td>I/II A</td>
<td>RT</td>
<td>Remission (12 months)</td>
</tr>
<tr>
<td>12</td>
<td>46/F</td>
<td>30 months</td>
<td>(ND)</td>
<td>(ND)</td>
<td>RT, HH-3</td>
<td>Relapses (16 months)</td>
</tr>
<tr>
<td>13</td>
<td>68/F</td>
<td>&gt; 2 years</td>
<td>CS</td>
<td>IV</td>
<td>TEM</td>
<td>Died (3 months)</td>
</tr>
<tr>
<td>14</td>
<td>66/F</td>
<td>Simultaneously</td>
<td>CS</td>
<td>IV</td>
<td>CS, TSPA</td>
<td>Died (11 months)</td>
</tr>
<tr>
<td>15,16</td>
<td>39/F</td>
<td>3 months</td>
<td>CS</td>
<td>(ND)</td>
<td>RT</td>
<td>Died (10 months)</td>
</tr>
<tr>
<td>17</td>
<td>36/M</td>
<td>5 months</td>
<td>CS</td>
<td>IVB</td>
<td>CS, chemo</td>
<td>Partial response (ND)</td>
</tr>
<tr>
<td>18</td>
<td>56/F</td>
<td>8 months</td>
<td>CS</td>
<td>IIIS</td>
<td>MOPP, RT</td>
<td>Remission (8 months)</td>
</tr>
<tr>
<td>19</td>
<td>29/M</td>
<td>5 years</td>
<td>CS</td>
<td>IV</td>
<td>RT, HH-3</td>
<td>Died (9 years)</td>
</tr>
<tr>
<td>20</td>
<td>71/M</td>
<td>13 months</td>
<td>(ND)</td>
<td>IIB</td>
<td>(ND)</td>
<td>(ND)</td>
</tr>
<tr>
<td>21</td>
<td>48/M</td>
<td>5.5 years</td>
<td>CS, MTX, CTX, Aza</td>
<td>IVB</td>
<td>MOPP, ABVD</td>
<td>Remission (12 months)</td>
</tr>
<tr>
<td>22</td>
<td>23/F</td>
<td>20 months</td>
<td>CS</td>
<td>IVA</td>
<td>MOPP, ABVD</td>
<td>Remission (4 months)</td>
</tr>
<tr>
<td>23</td>
<td>24/F</td>
<td>3 months</td>
<td>CS</td>
<td>II</td>
<td>Chemo</td>
<td>Died (ND)</td>
</tr>
<tr>
<td>24,25</td>
<td>65/F</td>
<td>1 year</td>
<td>CS, MTX</td>
<td>IVB</td>
<td>Withdrawal of MTX</td>
<td>Remission (1 year)</td>
</tr>
<tr>
<td>26</td>
<td>59/F</td>
<td>1 year</td>
<td>CS, IVIG, TAC, MMF</td>
<td>(ND)</td>
<td>ABVD</td>
<td>Died (2 months)</td>
</tr>
</tbody>
</table>

Index case 30/F Simultaneously CS IVB ABVD Improving (2 weeks)

On follow up, six patient died, eight patients (and the index case) showed response to the treatment (complete or partial), one patient relapsed and in one case there was no documented outcome. As sufficient follow-up of the patients and adequate documentation is not available, the prognosis of patients with HD and DM could not be determined.

Conclusion

Patients with DM have a definite increase in the risk of cancer. A high index of suspicion and a search for cancer at regular intervals is therefore required. Abnormal findings in the medical history and physical examination warrant an extensive search for occult neoplasm including screening with a PET scan and invasive procedures.

References