PHASE 1 ANCA-ASSOCIATED VASCULITIS WITH LUNG HEMATOMA

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Abstract

Anca-associated vasculitis is a group of systemic autoimmune diseases characterised by the presence of antibodies directed against proteinase 3 and myeloperoxidase. Lung haematoma is an uncommon presentation of ANCA-associated vasculitis. Here we present a case of a young female patient who presented with fever and pleuritic chest pain. The chest X-ray revealed a right sided pleural effusion with underlying lung haematoma. ANCA was positive and the patient was diagnosed with ANCA-associated vasculitis. She was treated with corticosteroid and immunosuppressant regimen. The patient had an excellent response to therapy and was discharged on follow up.

Key words: ANCA-associated vasculitis, lung haematoma

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was 31 years (range 24–64 years). All patients underwent breast ultrasound, mammogram, bone marrow biopsy and whole-body computed tomography for staging of lymphoma at presentation. Diagnosis was made on review of the blocks as all four patients had undergone lumpectomy at peripheral hospitals before being referred to our institute for further. Ann Arbor system was used for staging of lymphoma according to which bilateral cases were classified as Stage IVE due to poor prognosis and according to Wiseman and Liao criteria.[3]

At presentation, one patient has multiple bilateral skin nodules and two were diagnosed with pregnancy/lactation-associated breast lumps. Two of our patients had B symptoms and another two had bone marrow involvement at baseline. Histopathological diagnosis of DLBCL was made in three patients and one case had morphology consistent with mucosa-associated lymphoid tissue (MALT) lymphoma. Immunohistochemistry consistent with the given diagnosis was the positivity of CD 20, CD 30 and Ki 67 in three cases; however, immunohistochemistry was unknown for a single patient. There was no distant organ involvement at the time of presentation in all four cases.

The main stay of treatment was R-CHOP chemotherapy (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone). Surgery was limited for diagnostic purpose only, none of our patients received radiation therapy to breast. Response to the treatment was assessed by bone marrow biopsy after chemotherapy and PET scan. One patient died during the chemotherapy and the second patient had complete response, while rest of the two patients had progressive disease after chemotherapy, none of the patient received extensive surgery or radiation therapy, survival period ranges from 1 to 13 months; three patients died and two were lymphoma specific deaths [Table 2].

**Discussion**

PBL is a rare entity, it represents 0.04–0.5% of all breast malignancies.[6] Breast lymphomas are staged according to Ann Arbor staging system, although staging of bilateral PBL is still controversial, Ryan et al. classified bilateral PBL in Stage IVE,[5] this was the largest study of PB-DLBCL; they reviewed retrospectively 204 patients, whereas other studies include in Stage IE and IIE.[7,8] We included bilateral involvement in Stage IVE as they had poor prognosis. Bilateral PBLs have some association with pregnancy as mentioned in some studies.[9] It is difficult to differentiate radiologically between breast lymphoma, fibroadenoma and breast cancer.[1,10] The most common subtype is DLBCL accounts for about 50% of all BCLs, other rare types include MALT lymphoma, Burkitt lymphoma, anaplastic and follicular, small lymphocytic.[11] Aggressive form of PBL is known

### Table 1: Demographics

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Diagnosis</th>
<th>B symptoms</th>
<th>Pre-treatment bone marrow</th>
<th>Post-treatment bone marrow</th>
<th>Risks</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>27</td>
<td>DLBCL</td>
<td>No</td>
<td>Positive</td>
<td>Negative</td>
<td>Pregnancy associated</td>
</tr>
<tr>
<td>2</td>
<td>35</td>
<td>DLBCL</td>
<td>No</td>
<td>Negative</td>
<td>Negative</td>
<td>Multiple skin lesions</td>
</tr>
<tr>
<td>3</td>
<td>64</td>
<td>MALT lymphoma</td>
<td>Yes</td>
<td>Positive</td>
<td>Positive</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>24</td>
<td>DLBCL</td>
<td>Yes</td>
<td>Negative</td>
<td>Negative</td>
<td>Lactation associated</td>
</tr>
</tbody>
</table>

MALT: Mucosa-associated lymphoid tissue, DLBCL: Diffuse large B-cell lymphoma

### Table 2: Treatment and outcome

<table>
<thead>
<tr>
<th>Case</th>
<th>Chemotherapy</th>
<th>Radiotherapy</th>
<th>Response</th>
<th>Local relapse</th>
<th>Distant relapse</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>R-CHOP X 6</td>
<td>No</td>
<td>PR</td>
<td>No</td>
<td>No</td>
<td>Dead</td>
</tr>
<tr>
<td>2</td>
<td>R-CHOP X 6</td>
<td>No</td>
<td>CR</td>
<td>No</td>
<td>No</td>
<td>Dead</td>
</tr>
<tr>
<td>3</td>
<td>R-CHOP X 5</td>
<td>No</td>
<td>Progressive</td>
<td>No</td>
<td>No</td>
<td>Dead</td>
</tr>
<tr>
<td>4</td>
<td>R-CHOP X 4, RICE X 5</td>
<td>No</td>
<td>Progressive</td>
<td>No</td>
<td>No</td>
<td>Alive</td>
</tr>
</tbody>
</table>
to have association with younger age group; three of our patients were young.[12]

Another largest study by Jeanneret-Sozzi et al. included a review of 84 cases of PBL from 20 institutions, they concluded that PBL has better prognosis if managed by combined chemotherapy and radiotherapy.[13] MALT lymphoma breast is an extremely rare entity amongst the NHL, and bilateral involvement is even rarer, we found only single patient with bilateral MALT lymphoma.[14]

Chemotherapy stays the main treatment modality for breast lymphomas; R-CHOP is the preferred regime, as it increases 5-year survival. Aviv et al. recommended addition of Rituximab to chemotherapy may reduce central nervous system relapse and may be given intrathecal injections,[2,15] whereas other studies describe it as ineffective.[16] Radiotherapy can be combined with chemotherapy, but the role of surgery is limited only to diagnosis and palliation of symptoms in progressive disease. Jennings et al. observed high mortality rate in patients undergoing mastectomy and axillary dissection; therefore, they concluded that there is no role of axillary lymph nodal dissection and extensive surgery for breast lymphoma.[17,18]

Conclusion

Bilateral breast lymphoma is very rare, but aggressive disease with poor prognosis usually present at younger age group, aggressive treatment with chemotherapy should be considered and surgery should be reserved for diagnosis and palliation.

Acknowledgment

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Conflict of Interest

The authors declare that they have no conflict of interest.

References