Sole acquired trisomy 21 in a case of CD7 and CD10 positive acute myeloid leukemia

Sir,
A 52-year-old lady presented with fever and pallor since two months. She had bilateral cervical lymph node enlargement, gum hypertrophy, subconjunctival hemorrhage and sternal tenderness. Liver was enlarged 3 cm below costal margin. Spleen was not palpable. Her hemoglobin (Hb) 83g/L, total leukocyte count (TLC) 138.9 x 10^9/L and platelet count was 31 x 10^9/L. Peripheral smear showed 96% blasts. The bone marrow aspirate showed a hypercellular marrow with 96% blasts with auer rods. These blasts were myeloperoxidase, sudan black and non-specific esterase positive. These blasts were CD 33 and HLA DR positive. There was a co expression of CD7 and CD 10.

The 48h unstimulated culture-GTG banding demonstrated 47, XX, +21 in 100% cells examined [Figure 1]. There were no signs of Down’s syndrome in this lady.

Fluorescent in situ hybridization (FISH) done on buccal smears using the probe LSI 21 for 21q22.13-q22.2-region spectrum orange (Vysis, USA) showed that there was no aneuploidy of Chromosome 21 [Figure 2]. This was done to rule out congenital mosaic. FISH was negative for inv (16) (p13q22) & t (16; 16) (p13q22). Thus a final diagnosis of Acute myeloid leukemia (AML-M4) with sole acquired +21 was made.

Figure 1: Karyotype of the patient showing 47, XX, +21
She was given standard 3+7 chemotherapy. Her neutrophils recovered on day 27 and platelet counts recovered on day 37. A bone marrow aspirate done on day 36 showed her disease to be in remission. She was then given consolidation therapy with high dose cytarabine. Dose was not reduced and she tolerated it well.

Trisomy 21 (+21) in AML is usually present in conjunction with other cytogenetic changes, whose presence rather than +21 determines the clinical outcome. The incidence of +21 as a sole abnormality was between 0.3% in all patients with AML.\textsuperscript{[1-4]} Morphologically, AML with +21 as a sole abnormality preferentially shows M2 or M4 phenotypes according to the FAB classification.\textsuperscript{[1-4]} Cells trisomic for Chromosome 21 could be over-proliferating due to enhanced expression of a tumourigenic protein coded by a Chromosome 21 gene.\textsuperscript{[1]}

Expression of lymphoid antigens is common in AML. CD 2 is expressed in 16-21% and CD 19 in 7-14%.

Co expression of CD 7 on leukemic blasts has been documented in approximately 15% of AML.\textsuperscript{[2]} A high incidence of co-expression of CD 7 has been documented in the overall Down syndrome patients with leukemia. CD 10 has been occasionally reported in AML.\textsuperscript{[5]} The prognostic value of CD 7 and CD 10 expression in AML, however, is unclear. CD 7+ AML patients have a significantly lower response rate and poorer prognosis than CD 7- AML patients (Table 1).\textsuperscript{[5]}

Similarly, AML patients with acquired +21 as sole abnormality have been considered to have a poor prognosis.\textsuperscript{[1]} However, patients have shown CR and good prognostic indication.\textsuperscript{[2,3]} Co-expression of CD 7 is probably indicative of the very early stage at which the cell became malignant.\textsuperscript{[4]}

We conclude that sole acquired +21 with co-expression of CD 7 in AML is a rare phenomenon. Further data is required to assess the prognostic significance of CD 7 in this subgroup.

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age (years)</th>
<th>Sex</th>
<th>AML subtype</th>
<th>CD7</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wan et al.\textsuperscript{(1)}</td>
<td>28</td>
<td>M</td>
<td>M 2</td>
<td>NA</td>
<td>No treatment</td>
</tr>
<tr>
<td>Kondo et al.\textsuperscript{(2)}</td>
<td>21</td>
<td>F</td>
<td>M 2</td>
<td>+</td>
<td>CR, well at 4 months</td>
</tr>
<tr>
<td>Yamamoto et al.\textsuperscript{(3)}</td>
<td>49</td>
<td>M</td>
<td>M 2</td>
<td>+</td>
<td>CR, MDS 2 years later, leukemia 3 years later, died at 4 years</td>
</tr>
<tr>
<td>Udayakumar et al.\textsuperscript{(4)}</td>
<td>24</td>
<td>M</td>
<td>M 2</td>
<td>+</td>
<td>CR</td>
</tr>
<tr>
<td>Wei et al.\textsuperscript{(1)}</td>
<td>35</td>
<td>M</td>
<td>M 4</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Present case</td>
<td>52</td>
<td>F</td>
<td>M 4</td>
<td>+</td>
<td>CR, well at 2 months</td>
</tr>
</tbody>
</table>

\*M = Male; F = Female; NA = Not available; + = Positive; - = Negative; CR = Complete remission; MDS = Myelodysplastic syndrome

Figure 2: Fluorescent in situ hybridization on buccal smears for 21q22.13-q22.2-region shows no aneuploidy of chromosome 21

References

2. Kondo H, Kobayashi A, Iwasaki H. Trisomy 21 as the sole acquired karyotypic abnormality in an adult patient with CD7-positive acute

Sir,
The presentation and spectrum of skin cancer in Kashmir valley of Indian subcontinent is drastically different from rest of the country. Maxwell, in 1819 first reported skin cancer of the lower extremities in Kashmiri population attributing it to the use of Kangri.[1] Unlike skin cancers in general the biological behavior of these cancers is very aggressive with a substantial risk of loco-regional metastasis in 20-50% cases.[2,3] Surgery is the frequent modality of treatment used in the management of these tumors.

We want to share our experience with use of external beam radiotherapy alone in a case of recurrent Kangri cancer in the following index case. RT 377/06, a 46-year-old rural female developed itching followed by appearance of nodular swellings on the medial aspect of her right thigh over her previous operated scar (operated eight months earlier for her Kangri cancer). She was a chronic user of Kangri since her childhood. Her systemic examination was normal. Local examination revealed 3-4 nodular swellings with irregular margins, crusted surface and an indurated base over the previous operated site and in the background of reticular thermal keratotic lesions (erythema abignae) [Figure 1]. She refused re-do surgery or treatment with electrons however, agreed for an edge biopsy of the lesion which confirmed the diagnosis of squamous cell carcinoma with features of keratoses.

Patient was treated on a telecobalt unit and received external beam radiotherapy of 55 Gy/5 weeks by a direct portal with wax bolus [Figure 2]. She was also given prophylactic radiotherapy of 45 Gy/4 weeks to her inguino-femoral region. Patient completed the treatment protocol to both the sites without any toxicity and showed a complete response [Figure 3]. Patient is on regular follow-up and disease free at two and a half year follow-up.

A Kangri is an indigenous portable warming device in