Immunohematology Case Studies
2017 - 1

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Clinical History

• A 73 year old male
• Referred for redo vascular surgery on Jun 2016
• Osteomyelitis 2 years ago (May 2014)
• Antibody screen (ABID) was negative
• Transfused 3 occasions on Nov 2015 (a total 15 units during surgery)
• ABID positive (Jun 2016)
• Sample referred to Reference Laboratory after routine hours for investigation for following date surgery
• Patient is from Philippines & speaks Tagalog
Serologic and Transfusion History

Antibody screen negative on:
- 18 May 2014
- 05 Oct 2015
- 26 Nov 2015

Patient was transfused the following:
- 28 Nov 2015  9 units red cells transfused
- 23 Nov 2015  4 units red cells transfused
- 26 Nov 2015  2 units red cells transfused
Current Sample Presentation Data

- ABO/Rh: A, D+C+c+E+e+,K-
- DAT: negative
- Antibody Screen Method: using IH1000
- Antibody Screen Results: Panel reactive
- Antibody Identification Method: Bio-Rad IAT (3+), Bio-Rad enzyme IAT (4+) and Tube LISS-IAT (2+)

Antibody Identification Preliminary Results:
- Pan reactive suggestive of antibody to a high frequency antigen
Challenge with the Current Presentation

• Extended phenotyping was performed as this appeared to be antibody to a high prevalence antigen
• M+S+s+ excluded anti-U
• Lu(a-b+), Kp(a+b-) excluded anti-Lu^b or anti-Kp^b
• Fy(a+b-) excluded anti-Fy^3
• Jk(a-b-) suggestive the presence of anti-Jk3
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### Jk(a-b-) panel

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|---|---|---|---|---|---|----|---|---|-----|-----|-----|-----|-----|----|---|---|---|---|--------|----------|
| 1 | + | 0 | + | + | + | 0  | 0 | + | 0   | 0   | 0   | 0   | 0   | +  | + | + | + | 0 | +      | 0         |
| 2 | + | + | 0 | 0 | + | 0  | + | + | 0   | 0   | 0   | 0   | 0   | 0  | + | + | 0 | 0 | +      | 0         |
| 3 | 0 | 0 | + | 0 | + | 0  | 0 | + | 0   | 0   | 0   | 0   | 0   | 0  | + | + | + | + | +      | 0         |
Further Work

Testing for 2M Urea lysis

Jk(a-b-) cells lack the Urea Transporter (UT-B1) encoded by the SLC14A1 gene, and therefore are not lysed with 2M urea solution
Jk(a-b-) phenotype

- $Jk_{null}$ or Jk(a-b-) first reported by Pinkerton et al (1959) from a Filipino woman of Chinese & Spanish ancestry
- Cases of Jk(a-b-) are more frequent in the Polynesian & Finns
- Other populations, Chinese, Japanese, Asian Indians, Native Brazilians, African American, Tunisian, and European descent
Anti-Jk3

- Reacted optimally by IAT
- Enhanced by enzyme treated RBC
- Usually IgG, less common to be than IgM antibodies
- Complement binding
- Found in a non-transfused male
- No preference for Jk(a+b-) or Jk(a-b+)
- Not a mix of anti-Jk^a and anti-Jk^b
- No to severe/immediate or delayed transfusion reaction
- No to mild HDFN
- Auto anti-Jk3 has been reported
Transient Jk(a-b-) phenotype

Case report of a transient Jk(a-b-) phenotype

- Russian woman with myelofibrosis who made anti-Jk3 at the time her RBCs typed Jk(a-b-)
- Severe transfusion reaction
- Five weeks later typed as Jk(a+wb-)
- Anti-Jk^b was detected
- One year later typed as Jk(a+b-) with no anti-Jk3 and/or anti-Jk^b detected
Summary of Case Challenges

- Jk(a-b-) donations are rare
- Frozen and recovery donations were required for transfusion purpose
- Only anti-Jk3 was identified post transfusion in this case, using Jk(a-b-), Fy(a-b+) RBCs (exclusion of anti-Fy^b in the Fy(a-b+) patient)
- No confirmation of the presence of additional anti-Jk^a or anti-Jk^b
Lessons Learned by the Case

- Molecular basis of the Jk(a-b-) phenotype are diverse among the different populations.
- 2M Urea solution is considered easier and cheaper than genotyping to mass screen Jknull blood donors in countries with a significant prevalence of this phenotype.
- The ethnic origin and/or spoken language of the patient can give very important information about the putative rare blood type. In this case, the patient spoke Tagalog which is a Filipino dialect and quickly provided the clue for the antibody to a high prevalence antigen to be a likely anti-Jk3.
Molecular basis for JK phenotype

- **JK** gene ([SLC14A1], [HUT11A])
- Located at chromosome 18q12.3
- Jk\(^a\) antigen: p.Asp280 (c.838G)
- Jk\(^b\) antigen: p.As280 (c.838A)

**Carrier molecule**

Multi-pass glycoprotein.

*From Reid, Lomas-Francis & Olsson, The Blood Group Antigen Factsbook, 3rd Ed 2012*
Molecular based of some silencing of **JK*A or JK*B alleles**

Reference allele, **JK*02 (NM_015865)**, encodes Jk^b, Jk^3

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<th>Allele name</th>
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From Reid, Lomas-Francis & Olsson, The Blood Group Antigen Factsbook, 3rd Ed 2012
References


3. Ellisor SS, Reid ME, O’Day To et al. Autoantibodies mimicking anti-Jk\(^b\) plus anti-Jk\(^3\) associated with autoimmune haemolytic anemia in a primipara who delivered an unaffected infant. *Vox Sanguinis* 1983;45:53-59.


