Test Your Knowledge: Complement

NephMadness 2019 featured the **Complement Region**. The contemporary complement system lies at the interface of innate and adaptive immunity. Test your knowledge on the complement cascade and C3 glomerulopathy with the quiz* below.

1. Two sisters aged 14 and 16 years are diagnosed with C3GN. A mutation in the gene encoding which of the following is most likely?

    A. C3 Nephritic Factor
    B. Factor H
    C. C1
    D. MASP

2. A 60-year-old man with hypertension presents with new onset proteinuria, hematuria, and worsening kidney function. A kidney biopsy is performed that showed C3 glomerulonephritis. Which of the following is most likely to be an associated underlying disorder?

    A. Monoclonal gammopathy
    B. Staphylococcus aureus infection
    C. Systemic lupus erythematosus
    D. Diabetes mellitus

3. A 25-year-old female with aHUS causing ESRD undergoes a deceased donor kidney transplant. aHUS caused by a mutation in which of the following is LEAST likely to recur post- transplant?
    A. Factor H
    B. Factor I
    C. C3
    D. MCP

4. A 27-year-old man is diagnosed with aHUS and the decision is made to start eculizumab. He has received the standard Meningococcal vaccine. True or False: He should also be vaccinated against N. meningitidis serotype B.

    A. True
    B. False

- Quiz prepared by Anna Burgner, NephMadness Executive Team and AJKD Social Media Advisory Group Member. Follow her @anna_burgner.

To view the full Complement Region (FREE), please visit AJKDBlog.org.

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*This quiz was originally featured in the MOC Post-Test Questions for NephMadness 2019.

Answers to Test Your Knowledge: Complement

1. **B. Factor H**

   Of those listed, only Factor H mutations have been associated with genetic C3GN. C3 Nephritic Factor is an autoantibody that binds to and stabilize C3 convertase and is a cause of acquired C3GN, not genetic C3GN. C1 is part of the classical complement cascade, not the alternative complement cascade. MASP is part of the Lectin pathway.

   Reference:
   Dragon-Durey, M.A., Fremeaux-Bacchi, V., Loirat, C., et al. Heterozygous and homozygous factor H deficiencies associated with hemolytic uremic syndrome or membranoproliferative glomerulonephritis: report and genetic analysis of 16 cases. *JASN* 2004; 15(3): 787-95.

2. **A. Monoclonal gammopathy**

   The study referenced showed that 65% of patients older than 50 years with acquired C3GN had a monoclonal gammopathy present. The others are not known to be associated with C3GN.

   Reference:
   Ravindran, A., Fervenza, F.C., Smith, R.J., and Sethi, S. C3 glomerulopathy associated with monoclonal Ig is a distinct subtype. *Kidney Int.* 2018;94(1):178-86.

3. **D. MCP**

   As MCP is a membrane-bound protein and the allograft expresses normal membrane-bound, it is the least likely to cause recurrence of aHUS of those listed. The others listed are circulating factors and more likely to be associated with recurrence of aHUS.

   Reference:
   Zuber, J., Le Quintrec, M., Sberro-Soussan, R., Loirat, C., Frémeaux-Bacchi, V., and Legendre, C. New insights into postrenal transplant hemolytic uremic syndrome. *Nat Rev Nephrol* 2011; 7(1): 23-5.

4. **A. True**

   Patients receiving eculizumab are at high risk of developing N. Meningitis, even with receiving the standard vaccine. It is recommended that they are also vaccinated against serotype B of N. meningitidis.

   Reference:
   McNamara L.A., Topaz N., Wang X., Hariri S., Fox L., and MacNeil J. High risk for invasive meningococcal disease among patients receiving eculizumab (Soliris) despite receipt of meningococcal vaccine. *MMWR Morb Mortal Wkly Rep* 2017; 66(27): 734–7.

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