Screening and follow-up for paroxysmal nocturnal hemoglobinuria

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Introduction

PNH or Paroxysmal Nocturnal Hemoglobinuria is a rare hematopoietic stem cell disorder that is often suspected in a patient presenting with non-immune hemolytic anemia associated with pancytopenia or venous thrombosis. Screening of appropriate patients and accurate diagnosis of PNH are important.

Recently published paper (Mercier et al., 2017) investigated the clinical indications for PNH testing in the University hospital setting.

In this study we investigated the reasons for PNH analysis request and the results of the PNH tests in the non-University hospital setting, by using the methodology of Mercier et al.

Methods

We retrospectively reviewed flow cytometric PNH tests performed between February 2011 and February 2017. We determined the reason for PNH analysis request, based on the clinical and laboratory data.

We divided all requests into the following reasons for testing: hemolytic anemia (HA), aplastic anemia, cytopenia other than HA, thrombosis, MDS, follow-up (FU), no clinical data available.

Results

A total of 266 flow cytometric PNH tests (corresponding to 171 patients) were performed.

Of these 171 patients, 16 unique PNH-positive patients were identified, of which 8 had two or more PNH FU tests. 14 from the 16 unique PNH-positive patients were tested for PNH for the first time.

Table 1 shows the reasons for PNH testing in the 266 samples and in 90 PNH-positive samples, respectively.

Cytopenia (other than HA) was the most common reason for testing in all samples.

Reasons for testing	All samples N (%)	PNH-positive samples N (%)
Hemolytic anemia	29 (11)	4 (4,4)
Cytopenia other than HA	98 (36,8)	4 (4,4)
Aplastic anemia	6(2,3)	4 (4,4)
Trombosis	14 (5,3)	1 (1,1)
MDS	2 (0,75)	0
Follow Up	76 (28,6)	76 (84,4)
No clinical data	41 (15,4)	1 (1,1)

Table 1. Reasons for PNH testing.

86% of all samples and 96,6% of PNH – positive samples were requested by a hematologist (Table 2).

Specialisation	All samples N (%)	PNH-positive samples N (%)
Heamatology	228 (86)	87 (96,7)
Oncology	6 (2,3)	2 (2,3)
Gastro-Enterology	10 (3,8)	0
Cardiology	4 (1,5)	0
Nephrology	4 (1,5)	0
Internal Medicine	2 (0,75)	1 (1,1)
Geriatrics	3 (1,1)	0
Emergency	2 (0,75)	0
Other	7(2,6)	0

Table 2. Request for PNH testing per specialisation.

The highest probability of finding a PNH clone was founded in patients suffering from aplastic anemia (fig.1). Our findings in a non-university hospital setting is in agreement with the previous data in the university hospital setting of Mercier et al., (2017).

Figure 1. Probability to find a PNH clone

Conclusion

Cytopenia (other than HA) was the most common reason for testing in all samples.

The highest probability of finding a PNH clon was in patients tested because of aplastic anemia.

96,7% of PNH – positive samples were requested by a hematologist.

References

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