CORRESPONDENCE

Hemoglobin Henri Mondor [β 26(B8) Glu $\rightarrow$ Val, GAG $>$ GTG], A First Case Report in South-East Asia

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Dear Sir,

Hemoglobin (Hb) Henri Mondor [β 26 (B8) Glu $\rightarrow$ Val, GAG $>$ GTG; HBB:c.80A $>$ T] is a β -globin variant, at which codon 26 valine is substituted for glutamic acid and it is also a homologue of HbE [β 26 (B8) Glu $\rightarrow$ Lys, GAG $>$ AAG; HBB:c.79G $>$ A]. Hb Henri Mondor was first observed in a 9 years old African girl by Blouquit et al. in 1976 [1]. The second case was reported in an Algerian origin by Bardakdjian et al. in 1987 [2]. Here, we report for the first time, the Hb Henri Mondor in South-East Asia (SEA).

Case Report

A 20 years old Burmese man and his wife were seen by a physician at Lamphun Hospital, Thailand for an antenatal care. Their red blood cell indices for thalassemia screening were analyzed by using the automated blood counter (Sysmex KX-21, Sysmex Corporation, Kobe, Japan) and HbE screening was performed using dichlorophenol-indophenol (DCIP) test [3]. Test results are shown in Table 1. Both of them had microcytic hypochromic red cell

indices with low levels of MCV and MCH. In addition, the DCIP positive result was found in the husband's sample. Thus, their EDTA blood samples were sent to the Associated Medical Sciences–Clinical Service Center (AMS–CSC), Chiang Mai University, Chiang Mai, Thailand for further thalassemia and hemoglobinopathy diagnosis. The Hb analysis was performed using high performance liquid chromatography (HPLC; VARIANT IITM, β -Thalassemia Short Program; Bio-Rad Laboratories, Hercules, CA, USA). The results are shown in Table 1. The woman's sample showed normal HPLC chromatogram while the man's sample showed abnormal Hb peak with a level of

Table 1 Characteristics and red blood cell parameters of the Burmese couple

Characteristic	Man	Woman
Age (years)	20	19
Red blood cell counts ($10^{12}/L$)	5.5	4.0
Total Hb (g/dL)	14.0	10.6
PCV (L/L)	0.42	0.31
MCV (fL)	75.8	77.2
MCH (pg)	25.5	26.8
MCHC (g/L)	333	342
RDW (%)	13.4	13.9
DCIP test	Positive	Negative
Hb A (%)	57.4	91.0
Hb A ₂ (%)	2.0	2.8
Hb F (%)	0.3	0.0
Hb Henri Mondor (%)	30.7	–
α -Globin genotype	$\alpha\alpha/\alpha\alpha$	$\alpha\alpha/\alpha\alpha$
β -Globin genotype	$\beta^A/\beta^{CD26(B8) \text{ Glu} \rightarrow \text{Val, GAG} > \text{GTG}}$	β^A/β^A

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Fig. 1 The HPLC chromatogram (a), CE electrophoregram (b) and DNA sequence (c) of the man with Hb Hb Henri Mondor

30.7% at the retention time of 2.86 min (Fig. 1a). His blood sample was then analyzed for the hemoglobin analysis by capillary electrophoresis (CE) (CapillariesTM 2 Flex Piercing; Sebia, Norcross, GA, USA). An abnormal Hb peak with a level of 34.5% was observed at the electrophoretic zone D with a migration time of 195–210 s (Fig. 1b). The molecular analysis yielded negative results

for α -thalassaemia-1 (SEA and Thai type deletions) and α -thalassaemia-2 ($-\alpha^{3.7}$ and $-\alpha^{4.2}$ kb deletions). In addition, the PCR amplification of the β -globin gene was accomplished and then the direct DNA sequencing of the amplified product was performed on an ABI PRISMTM 3130 XL analyzer (Applied Biosystems, Foster City, CA, USA). The results showed molecular defect causing

abnormal hemoglobin resulting in A to T transversion at the second position of codon 26 (GAG > GTG) of β -globin gene (Fig. 1c) that leads to a substitution of valine for glutamic acid described previously as Hb Henri Mondor [1].

Discussion

The amino acid substitution present in Hb Henri Mondor β 26 (B8) Glu $\rightarrow$ Val occurs in the same position as that of HbE β 26 (B8) Glu $\rightarrow$ Lys [1]. This substitution involves an external residue, located at a distance from the major functional domain of the molecule. Thus, it exhibits a normal oxygen affinity which is similar to HbE [2, 4]. In addition, the hemolysate containing Hb Henri Mondor showed a slightly increased hemoglobin precipitation as did the hemolysate containing HbE, and thus, it can be detected by DCIP test [2]. However, a slight molecular instability did not produce a hemolytic syndrome. In current study the man with heterozygous for Hb Henri Mondor had low levels of MCV and MCH while his red blood cell counts, total Hb, PCV, MCHC, and RDW were observed within the reference ranges. However, his serum iron and serum ferritin were not measured.

In conclusion, Hb Henri Mondor can be found in the South-East Asian population. Therefore, better

understanding of HPLC chromatogram and/or CE electrophoregram patterns and hematological and clinical features of this Hb variant will be useful for genetic counseling.

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