Pterygium in the patients with hemoglobin E containing diseases: The preliminary report

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Abstract: This paper is aimed to find the prevalence of pterygium among the patients who had hemoglobin E containing diseases. **Patients and Methods:** The cross-sectional study was conducted among the patients with Hb E trait, Hb E disease, Hb AEBart, Hb CS-AEBart, beta-thalassemia/Hb E, Hb EFBart and Hb CS-EFBart diseases. They were examined for the presence of pterygium. **Results:** From 124 cases, pterygia had been observed in 28 cases (22.6 %), 12 cases having pterygia in both eyes (9.7%) and 16 cases having pterygium in only 1 eye (12.9%), 10 in the right eye and 6 in the left eye. The severity of pterygia consisted of grade 1 of 34 eyes, grade 2 of 4 eyes and grade 3 of 2 eyes. **Conclusion:** The prevalence of pterygium in the patients with hemoglobin E containing diseases is rather high. It cannot be solely explained by the sun exposure alone. However the true association needs further investigations.

บทคัดย่อ: ต้อเนื้อในผู้ป่วยที่มี ฮีโมโกลบิน อี การศึกษาเบื้องต้น

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วัตถุประสงค์: ศึกษาหาความชุกของโรคต้อเนื้อในผู้ป่วยที่มีฮีโมโกลบิน อี **ผู้ป่วยและวิธีการ:** เป็นการศึกษาภาคตัดขวางในผู้ป่วยที่มีฮีโมโกลบิน อี ได้แก่ผู้ป่วย Hb E trait, Hb E disease, Hb AEBart, Hb CS-AEBart, beta-thalassemia/Hb E, Hb EFBart and Hb CS-EFBart diseases ว่ามีต้อเนื้อหรือไม่ **ผลการศึกษา:** มีผู้ป่วย 124 ราย พบมีต้อเนื้อ 28 ราย (ร้อยละ 22.6) โดย 12 รายมีต้อในตา 2 ข้าง (ร้อยละ 9.7) และ 16 รายมีต้อเพียงข้างเดียว(ร้อยละ 12.9), พบต้อในตาขวา 10 คน และในตาซ้าย 6 คน ความรุนแรงของต้อพบว่า grade 1 มี 34 ตา, grade 2 มี 4 ตา grade 3 มี 2 ตา **สรุป:** ต้อเนื้อพบมากในผู้ป่วยที่มีฮีโมโกลบิน อี อาจจะเป็นการพบร่วมโดยบังเอิญ หรืออาจจะมีความเกี่ยวข้องกันจริง ยังต้องการการพิสูจน์ในการศึกษาต่อไป

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Background

Hemoglobin E (Hb E) is the genetic abnormality resulted from the combination of 2 alpha and 2 abnormal beta globin chains, i.e. glutamine at the 26th position of the beta chain is substituted by lysine. It combines with other thalassemias and/or hemoglobinopathies to form heterogeneous genetic complexes, resulting in different clinical severity. Hb E trait is always asymptomatic while Hb E disease may have mild degree of microcytic anemia and mild splenomegaly[1]. In other forms, i.e. Hb AEBart, Hb Constant-Spring (CS)-AEBart, beta-thalassemia/Hb E, Hb EFBart and Hb CS-EFBart diseases, the patients have some degree of hepatospleno-megaly and hemolytic anemia. All these Hb E containing diseases are highly prevalent in Thailand, e.g. 30-50% in the northeastern part of the country[2,3].

Pterygium is the benign fleshy growth protruding from limbus onto the cornea. It can cause mild irritation and lacrimation. The etiologies are not known but it is more common in the tropical zone, mostly related to dose-dependent sun exposure[4] while the genetic back ground may play some roles[5]. Thailand is in the tropical zone and her northeastern part is sunnier, drier and higher. Therefore pterygium is expected to be more frequent.

This study is aimed to report the prevalence of pterygium in the patients who carry Hb E with/without alpha, beta-thalassemia and/or other hemoglobinopathies.

Patients and Methods

This cross-sectional study was conducted upon the patients with Hb E containing diseases, i.e. Hb E trait, Hb E disease, Hb AEBart, Hb AEBart-CS, Hb EFBart and Hb EFBart-CS diseases. All were diagnosed by clinical manifestation, hematological parameters and Hb electrophoresis. They all regularly attended hematology clinic at Maharat Nakhon Ratchasima Hospital.

They were examined to have pterygium from September to December of 2010. The severity of the pterygium would be classified into 3 grades, i.e. graded 1, if the length of pterygium was less than the half of the distance between the limbus and the pupil, grade 2 if its length was more than the half of such the distance, grade 3 if the pterygium crossed into the pupil[6].

Pinguecula was not included in this study. The age of the patients included was 15 years old or more. The patients with phthisis bulbi were excluded.

Results

During 3-month period of the study, 124 patients who had Hb E-containing diseases were recruited. They consisted of 32 males and 92 females, ages ranged between 15 to 77 years, mean of 42.3+14.1 years. The mean of hemoglobin concentration was 10.0+2.8 g/dL. The kind of Hb E containing diseases was shown in the table 1.

Table 1 The various kinds of Hb E-containing diseases

Kinds of Hb E-containing diseases	Cases (%) (N = 124)
Hb E trait	48 (38.7)
Hb E disease	33 (26.6)
Beta-thal/Hb E disease	28 (22.6)
Hb AEBart-CS disease	8 (6.4)
Hb EFBart disease	4 (3.2)
Hb AEBart disease	3 (2.4)

From 124 cases, pterygia had been observed in 28 cases (22.6 %), 12 cases had pterygia in both eyes (9.7%) while 16 cases had in only 1 eye (12.9%). Most of them had pterygium at the medial side.

For the patients who had the pterygium in the only one eye, 10 in the right eye and 6 in the left eye. The severity of pterygia consisted of grade 1 of 34 eyes, grade 2 of 4 eyes and grade 3 of 2 eyes. Each patient who had pterygia in both eyes had the same grades between both eyes. All of them were recommended to consult the ophthalmologists for the proper management.

Discussion

The overall prevalence of pterygium among 124 patients with Hb E containing diseases was 22.6%, and the majority of them were in grade 1.

In the population based study of adults aged 21 years in Riau province of Indonesia, an area near to the equator, the age adjusted prevalence rate of any pterygium was 10.0% and of bilateral pterygia was 4.1%[7]. In the population base survey in Singapore, the prevalence of pterygium among Chinese aged 40 years and older was 7%[8]. These prevalences are lower than that of our study even these studies were conducted in the countries which are nearer to the equator than Thailand. It sounds that the proximity to the equator is not the sole factor for the development of pterygium.

Under the same environment in the very small country like Singapore, there were different prevalences of pterygium between the different ethnic groups, i.e. 12.3% among Malays[9] and 7% among Chineses[8]. From this point of view, the genetic basis itself or other related factors may play some important roles for the development of the pterygium.

In the prospective cross-sectional, case-control study in Nigeria, the prevalence rate of pterygium in commercial motorcyclists was 12.5% compared with 7.9% of the control group, they are lower than ours. It suggests that the outdoor activity is not the only one factor that promotes the development of pterygium[10]. In the 9-year study in Barbados, an average incidence of pterygium is 1.3% per year whereas the 9-year incidence is 11.6%. Working outdoors increased the risk 1.5 fold[11]. This work still emphasizes the outdoor activity as the important associated factor for the pterygium.

The study in Myanmar showed the prevalence of pterygium was 19.6% for 1 eye and 8% for 2 eyes, and it was found that outdoor occupation was an independent predictor of pterygium[12]. It is higher than ours, presumably due to the age of the studied population which was started at 40 years or more years and the studied area which focused on rural.

The study of Gazzard et al[7] showed the importance of the increasing age for the increasing prevalence of pterygium in Indonesia, i.e. 2.9% for 21-29 years vs. 17.3% for 50 years or above.

Besides the known factors relating to the development of pterygium such as ultraviolet exposure, the increasing age and outdoor work[13], one associated factor was the urokinase plasminogen activator (uPA) which was the protease involved in tissue remodeling and cell migration. Chao et al found the overexpression of uPA in the pterygium and their fibroblast, it was presumed to convert plasminogen to plasmin, degrade extra-cellular matrixes, stimulate cell migration, induce angiogenesis and play an important role in the development and progression of pterygium[14].

In the further studies, this interesting fact should be verified in the cases of hemoglobin E containing diseases.

Conclusion

The prevalence of pterygium in the patients with hemoglobin E containing diseases is rather common in this study. It cannot be solely explained by the sun exposure alone. However the true association needs further investigations to compare the prevalence of pterygium between the group of hemoglobin E containing diseases and the normal control.

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