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Harris Platelet Syndrome: The Need to Recognise the Entity.

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Short Communication

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ABSTRACT

Inherited giant platelet disorders (IGPD), is a subgroup of congenital thrombocytopenias, and is characterized by decreased platelet counts along with macro platelets and variable bleeding symptoms. Harris Platelet Syndrome a newly described entity, previously called asymptomatic constitutional macrothrombocytopenia is a sub-type of IGPD and is characterized by mild to severe thrombocytopenia, macroplatelets and no bleeding manifestations in the North Western part of Indian sub-continent. We report, the occurrence of Harris platelet syndrome in a group of population from northern part of West Bengal (part of North Western Indian sub-continent geographic location).

INTRODUCTION

Congenital thrombocytopenias are an extremely rare group of haematological disorder [1]. However, Inherited giant platelet disorders (IGPD), a subgroup of congenital thrombocytopenia, characterized by decreased platelet counts along with macro platelets and variable bleeding symptoms, once considered a very rare entity is now detected with increasing frequency [2]. Currently, IGPD's are classified into 4 broad sub-categories: the first category is based on structural defects of platelets; the second category is based on pathognomonic peripheral blood morphologic findings like neutrophil inclusions; the third category is based on associations with clinical and systemic manifestations; the fourth category is considered a benign anomaly such as Mediterranean macrothrombocytopenia[3]. Very recently, another cohort of population from north eastern part of Indian sub-continent, were found to have Harris platelet syndrome or asymptomatic constitutional macrothrombocytopenia[4]. This recently described syndrome by Naina et al [4], is characterized by mild to severe thrombocytopenia, macroplatelets and no bleeding manifestations. We report, the occurrence of Harris platelet syndrome in a group of population from northern part of West Bengal, India who had presented for pre-employment health check-up.

MATERIALS AND METHODS

CASE REPORT

During May, 2013, 60 male subjects were screened, during their pre-employment health check-up. Among them 31 were from northern part of West Bengal and 29 were from Tamil Nadu. Blood samples were collected in di-potassium ethyl-enediaminetraaceitic acid (EDTA) vaccutainer. Automated blood analysis including platelet analysis was performed using automatic blood analyzer (Beckman Coulter Act 5 Diff) within 3 hours of collection, as part of their pre-employment health screening. Peripheral blood smears stained with leishman stain were examined to confirm the presence of thrombocytopenia and giant platelets. Statistical evaluation was performed by statistical package for the social sciences (SPSS) version 16 program using Student's independent sample two-tailed t-test. A p value <0.05 was considered statistically significant.

Platelets less than 150 * 10^9/Lwere categorized as thrombocytopenia. Further, thrombocytopenia were sub-categorized according to the platelet counts into mild thrombocytopenia (100 to 150 * 10^9/L), moderate thrombocytopenia (50 to 100 * 10^9/L) and severe thrombocytopenia (<50 * 10^9/L). Platelets with mean platelet

volume greater than 10 fl were labelled as macroplatelets. Twenty one (35%) West Bengal subjects had thrombocytopenia; amongst them eleven had mild thrombocytopenia (18.3%), ten had moderate thrombocytopenia (16.6 %) while none of them had severe thrombocytopenia.

OBSERVATIONS AND RESULTS

All West Bengal subjects with thrombocytopenia also had macro-platelets. On the other hand, no subjects from Tamil Nadu had thrombocytopenia or macroplatelets (Table 1).

Table 1: Summary of platelet count, mean platelet volume in subjects from West Bengal and Tamil Nadu

COHORT	NUMBER OF SUBJECTS	PLATELET COUNT [* 10^9/L]			PLATELET VOLUME[fL]		
		MEAN	RANGE	SD	MEAN	RANGE	SD
ALL SUBJECTS	60	204	54 to 395	92.76	9.74	6.4 to 14.1	2.45
WEST BENGAL	29	124	54 to 210	44.86	12.06	9.3 to 14.1	1.09
TAMIL NADU	31	278	169 to 395	56.14	7.57	6.4 to 10.2	0.80

Table 2: Summary of difference in platelet count and platelet volume between subjects from West Bengal and Tamil Nadu

PARAMETER	MEAN DIFFERENCE	95% CONFIDENCE INTERVAL		p value
		Lower	Upper	
PLATELET COUNT	-154	-180	-127	< 0.01
PLATELET VOLUME	4.5	4	4.9	< 0.01

The West Bengal subjects had significantly lower platelet count (mean difference 154*10^9) when compared to Tamil Nadu subjects. Moreover, the platelet volume was significantly higher (mean difference 4.5fl) among West Bengal subjects when compared to Tamil Nadu subjects (Table 2).

Subsequently, a detailed clinical, therapeutic history and clinical examination was done to rule out other inherited thrombocytopenias and immune thrombocytopenias. There was no history of excessive bleeding; no significant systemic/clinical findings; and no significant morphologic findings in the peripheral blood smear.

DISCUSSION

The literature search of this peculiar finding in a specific cohort revealed the presence of new entity named Harris Platelet syndrome, previously labelled asymptomatic constitutional macrothrombocytopenia in north eastern part of the Indian sub-continent [2, 4]. Echoing the findings of Naina et al[4], we found a cohort from northern part of West Bengal with moderate to mild thrombocytopenia and macroplatelets, no clinical or bleeding manifestations and absent significant peripheral blood morphologic findings. We strongly believe this cohort from West Bengal; also belong to the cohort of Harris platelet syndrome. However, there are no confirmatory tests to diagnose this entity. It still remains a diagnosis by exclusion of other IGPD's. We believe this entity, Harris platelet syndrome belongs to the fourth category of IGPD (A benign anomaly) and as similar benign course as Mediterranean macrothombocytopenia. A proper confirmatory test, clinical outcome and course of this entity should be elicited in order to get a proper understanding of this entity. With the advent of global migration, it is no longer a locus specific entity. Hence a proper understanding of this entity is the need of the hour to alleviate unnecessary anxiety, treatment to the subjects with Harris platelet syndrome in any part of the world.

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