Benign hematological disorders in India: The status

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Hematological disorders notwithstanding, the load of benign hematological disorders in India is alarming. Benign hematological disorders can be broadly divided into those with acquired and inherited causes. Acquired causes due to iron and vitamin deficiency indeed exceed the congenital causes. These are more common in women than in men. Women get iron deficiency more often due to excessive bleeding during menstruation and child birth. Worm infestation and piles contribute to iron deficiency anemia both among men and women. Occult malignancies which cause bleeding are also well-known causes for iron deficiency anemia. Nutritional deficiency, multiple pregnancies, and malabsorption contribute to dimorphic anemia—a combination of iron and folate/vitamin B12 deficiency.

In a hospital-based study in Delhi, it was observed that 2.7% of all anemias were megaloblastic anemia. In this study of 175 patients, majority of patients had lone vitamin B12 deficiency, which was followed by combined deficiencies of vitamin B12 and folate. Only folate deficiency was observed in a very small number of patients. An interesting observation was the use of acid-suppressing medication contributing to megaloblastic anemia in one in six patients.[1]

More and more reports from India now show vitamin B12 deficiency as a predominant cause for megaloblastic anemia in contrast to folate deficiency, as earlier believed. This may be attributed to the routine folic acid supplementation, especially among women now.[2]

Western data emphasizes on pernicious anemia as the major cause, but Indian data suggests nutritional deficiency of vitamin B12 and folic acid as the major cause of megaloblastic anemia.

In a study at Pune, Maharashtra, it was observed that three-fourths of a community had biochemical proof of vitamin B12 deficiency in their blood, though only a small part of this population was vegetarian.[3]

In another study from Puducherry, megaloblastic anemia was found to be present in 38.4% of 60 adult patients of macrocytic anemia. The megaloblastic anemia observed was due to cobalamine deficiency in majority of cases; few had combination of cobalamine and folate deficiency. None had lone folate deficiency. A significant observation was that only one-fourth of this group was vegetarian.[4]

Diseases like chronic renal failure, aplastic anemia, anemia of chronic disease, and hemolytic anemias do contribute to the load of anemia.

One of the important causes for inherited hemoglobinopathies in India is thalassemia. The reasons proposed for high incidence of thalassemia are:

1. Being an equatorial population, the genes that cause thalassemia and sickle cell disease developed as a defense mechanism against malaria among some tribes and communities. This could be due to an evolutionary gene mutation.

2. The other reason put forth is that the Greeks, among whom thalassemia was widespread, had mixed with the local populations during Aryan invasion centuries ago and had given them the disease. Since the mixing happened primarily with the people of North India, it is mostly seen among Aryans in North India and is negligible among Dravidians in the South.

A high incidence of alpha-thalassemia, beta-thalassemia, and Hb variants has also been observed among Gonda tribes in Madhya Pradesh in central India.[5] Another blood disorder that is common among certain ethnic groups from North India is glucose-6-phosphate dehydrogenase (G6PD) deficiency.

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The highest incidence of thalassemia in the world is in Maldives with a carrier rate of 18% in the population. The prevalence is 16% among the people from Cyprus. Overall prevalence in India is 3-8%; however, one in eight persons in India is believed to be carrier of the thalassemia gene. Routine screening, as followed in Cyprus and Iran, should be implemented in India at highly prevalent regions like Punjab, West Bengal, and northeastern states.

Thalassemia co-exists with other hemoglobinopathies also in India. The most common of these are:
1. Hemoglobin E/thalassemia: Common in eastern parts of India
2. Hemoglobin S/thalassemia: Common in western Orissa
3. Hemoglobin D/thalassemia: Common in north-west part of India which includes Punjab region

Another hemoglobinopathy found at a menacing proportion in certain parts of India is sickle cell disorder. It has been observed in about 34% of Gujarat’s population.[6] In Maharashtra also, there is a very high incidence of sickle cell anemia.

Western Orissa too has a high number of sickle cell disease patients. Approximately 1 out of 10 people has this disease in this region.[6] Besides, mixed hemoglobinopathies like sickle-thalassemias are also found to be quite common in this region. In one study, 8.1% of the population had sickle-beta thalassemia gene.[6] Consanguinity may also be one of the contributing factors for the high incidence.[6]

In a study of abnormal hemostatic function, 630 of 768 patients were diagnosed to have hereditary bleeding diathesis at Indian Institute of Immunohematology, Mumbai. Only 5% of these patients had platelet dysfunction, the most common being Glanzmann’s thrombasthenia followed by Bernard-Soulier syndrome.[9] The remaining patients had coagulation dysfunction. Hemophilia A was the commonest coagulation disorder found, followed by hemophilia B and von Willebrand disease.

In a similar analysis of genetic bleeding disorders from Sanjay Gandhi Postgraduate Institute, Lucknow, hemophilias A and B were found to be the commonest bleeding disorders, followed by von Willebrand disease and inherited thrombocytopenias.[10]

In a study of 337 women with inherited bleeding disorder who presented with menorrhagia, isolated PF3 availability defect and von Willebrand disease were found to be the commonest causes.[11] In another retrospective analysis of clinical profile of 67 patients with rare inherited coagulation disorders from the same center, factor X deficiency was found to be the commonest defect.[12]

In view of the high prevalence of bleeding disorders, it is desirable that infrastructure should be widened at district hospitals to detect the bleeding disorders. Even in all first-degree female relatives of severe and moderate hemophilia patients, factor assays should be performed because some of them may be vulnerable to post-partal or post-traumatic bleeding. Pre-natal diagnosis with chorionic villus sampling, amniocentesis, and fetal blood sampling should be done in appropriate cases in early pregnancy.

Overall, the major burden of benign hematological disorder consists of nutritional anemias. Adequate supplementation of iron and vitamins to the vulnerable group will go a long way to contain this problem.

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