Volume : 3 | Issue : 8 | Aug 2014 • ISSN No 2277 - 8179

Research Paper

Medical Science

KEYWORDS : Piebaldism, White forelock, Genetic counseling

PIEBALDISM: a Rare Melanocytic Disorder

Moiz F. Mithaiwala M.D. (Skin & V.D.), consultant dermatologist, general hospital, Jamkhambhaliya.

Alka K. Raka M.D. (Skin & V.D.), consultant dermatologist, G.G.hospital, Jamnagar.

ABSTRACT

Piebaldism is a rare autosomal dominant disorder of melanocytic development, characterized by frontal, median or paramedian depigmented macules and patches with white forelock. We report the case of 10 year male child with typical features of piebaldism with positive family h/o white forelock present. All vitals are stable with routine investigation in normal limit. There was absence of melanocytes and melanin in depigmented areas and hair bulbs. Genetic counseling, photo protective measures and static nature of disease was explained.

INTRODUCTION:

Piebaldism is rare autosomal dominant disorder of melanocytic development which is known since Egyptian times. Roman Generation after generation demonstrated a distinctive predictable family mark – white forelock. Many family known by this name of white forelock, holilock etc. This case report discusses a unique presentation of this disorder in 10 year male child with family h/o positive.

CASE REPORT:

A 10 year male child patient presented in SKIN O.P.D. with asymptomatic white colour patches with white colour hair at center on of forehead. Lesion was static in nature. Patient did not had any complain of burning, itching. No other c/o regarding any other system. In family history, father have white forelock present.

On examination, pt had vitals stable. Multiple, depigmented, symmetrical patches which mainly involved anterior part of chest, middle part of upper arm, lower limb were present. White forelock with similar patch over middle part of forehead with leukotrichia of eyebrow and eyelash. Back and acral part were relatively spared. There are islands of normal skin in amelanotic skin. On Woods lamp examination it shows pearly white colour fluorescence. ENT and Ophthalmic examination does not show any abnormal significant finding.

DIFFERENTIAL DIAGNOSIS:

included Waardenburgs syndrome, Vitiligo, Tinea versicolor, and Leprosy.

Patient suffering from Waardenburg syndrome presented with increase in interpupillary distance, heterochromia, sensorineural hearing loss which was not present in our patient. As Vitiligo is progressive in nature which in contrast to our case, in which lesion present since birth and static in nature. Tinea versicolor is ruled out by Woods lamp examination as there is no yellow green fluorescence and KOH examination ,which was negative in our patient. Our present case had sensation normal in skin lesion and at periphery. Also there is no peripheral nerve thickening present, so leprosy can be ruled out.

It was likely that patient was suffering from rare disorder of Piebaldism.

DISCUSSION:

Piebaldism is autosomal dominant disorder of pigmentation characterized by white hair and skin with lack of melanocytes. The world piebald itself has been attributed to a combination of 'Pie' in magpie (bird of black & white plumage) & the "bald" is of bald eagle (The United States national bird, which has a white featherhead). Piebaldism is benign disorder. Incidence of piebaldism is estimated to lower than 1 in 20,000(1). There are very few Indian reports of this disorder (2, 3).Piebaldism caused by mutation in KIT proto-oncogene on chromosome 14(4).

Melanocytes and melanin are not present in the hair bulbs and white patches of the depigmented areas. Dopa reaction is not positive in the depigmented skin. In the hyper pigmented macules, melanocytes are detected in normal numbers with abundant melansomes.The differentiation, migration and perhaps the survival of melanoblasts are at fault. The distribution of patches may be explained on the basis of impaired migration of melanoblast, so that areas near their origin are spared while that failed to arrive at the most distant sites (5).

Treatment option which can be given- Genetic couselling, Photoprotection (6), Dermoabrasion, split skin grafting followed by minigrafting -Autologous melanocytic transplantte (7). PUVA & topical corticosteroid does not give promising result.



Figure 1: Face of the child showing white forelock with white patches

Volume : 3 | Issue : 8 | Aug 2014 • ISSN No 2277 - 8179



Figure 2: Front of the child showing multiple depigmented patches with islets of hyperpigmentation



Figure 3: Back of the same child showing multiple patches on hands and legs

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