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Case Report

Carbon baby syndrome: two case reports

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Abstract Carbon baby syndrome, also known as universal acquired melanosis is one of the causes of diffuse hyperpigmentation of skin and mucosa with only a limited number of reported cases in the literature. We here report, two cases of universal acquired melanosis in two siblings of a family. They developed progressive darkening of skin and mucosa from 5 and 4 months of their lives, respectively. The colour change was insidious, asymptomatic, progressive and generalised. Histopathology showed increased pigmentation of epidermal basal layer. According to clinicohistological assessment, the cases were diagnosed as carbon baby syndrome with familial clustering.

Key words
Acquired, diffuse hyperpigmentation, familial clustering, carbon baby.

Introduction

Acquired universal melanosis, an extremely rare condition is characterised by progressive pigmentation of the skin during childhood, resembling that seen in black races. The major determinant of normal skin colour is the melanin, produced by melanocytes of basal layer of epidermis. Melanin production normally is not maximal in the newborn skin. Baby skin is slightly tanned for few days after birth and it turns into racial colour after few months due to repeated sun exposure. Our two patients presented with diffuse hyperpigmentation which was progressive from the ages of 4 and 5 months of their lives, respectively. To the best of our knowledge, carbon baby syndrome in two siblings of a family has not been described previously. We, therefore, report these two cases here.

Case report 1

A 5-year-old male child born of consanguineous marriage was brought to the dermatology department with complaints of progressive darkening of whole body. His mother had uneventful prenatal, natal and postnatal periods. The pigmentation started in the face and hands at the age of 5 months and then gradually progressed over the next one year to involve the whole body. There was no history of fever, skin infection, darkening of urine, photosensitivity or any other systemic complaints. History of any prior drug intake was absent. Family history was positive. His 3-year-old female sibling was affected by the same disorder of generalised acquired hyperpigmentation.

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Case report 2

A 3-year-old female child born of consanguineous marriage, presented with progressive darkening of whole body. History revealed uneventful prenatal, natal and postnatal periods. History of long-term drug intake by the mother was absent. The increased pigmentation was first noticed over the face at the age of 4 months and then gradually involved the entire body. There was no history of fever, skin infection, darkening of urine, photosensitivity, any other systemic

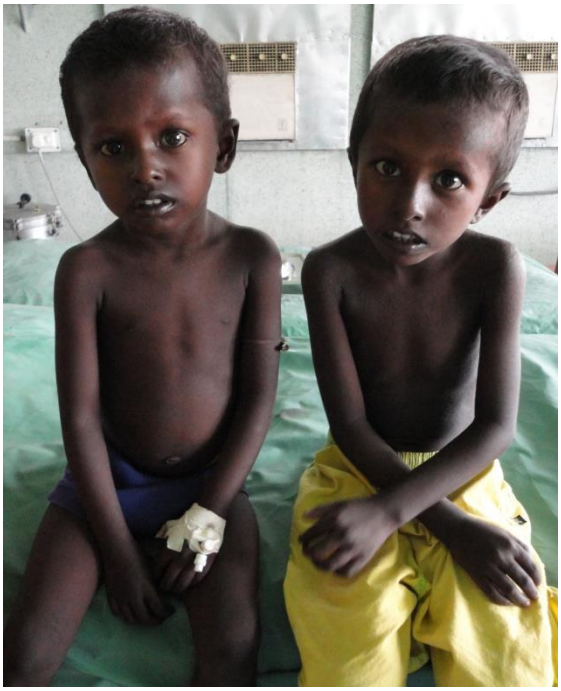

Figure 1 Two siblings with diffuse hyperpigmentation.

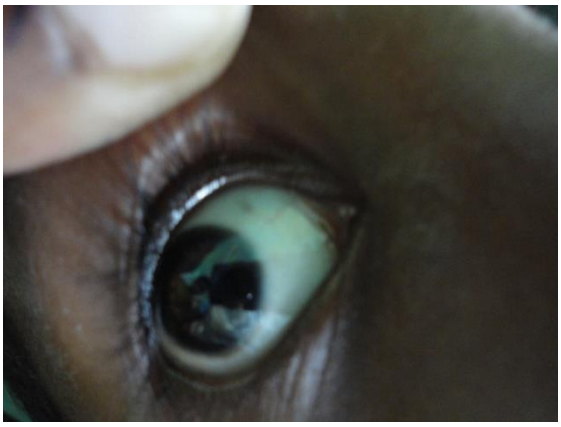

Figure 2 Conjunctival patch (mucosal pigmentation).

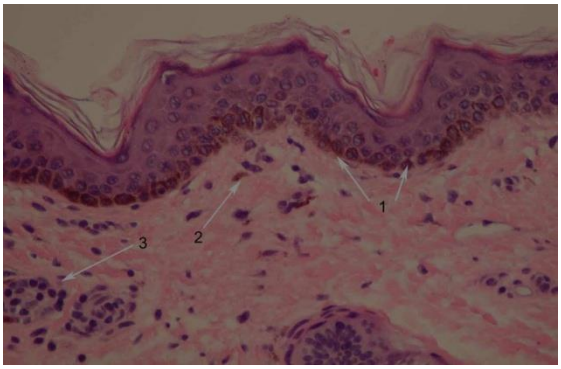

Figure 3 Hematoxylin and eosin staining showing increased pigmentation of basal (mainly) and suprabasal layers with some dermal melanophages.

complaints or prior medication. Family history was positive. Her 5-year-old male sibling was affected by same disorder of generalised acquired hyperpigmentation.

On examination, both of them had generalised diffuse hyperpigmentation of entire body (**Figure 1**). Dark patches were seen over upper bulbar conjunctiva (**Figure 2**), oral mucosa and tongue. Palms and soles were also involved. No abnormality of hair and nails was detected. Hyperpigmentation was more pronounced over sun-exposed areas. Growth parameter and developmental milestones were within normal limits. No other system was involved. On ophthalmoscopic examination, retina and fundus were normal. Routine blood and urine examinations were within normal range. No abnormalities were detected in liver function tests and thyroid function tests. Serum corticotropin was in normal range and there was no diurnal variation. Histopathology of skin showed increased melanin pigmentation in basal and suprabasal layers of epidermis and a few melanophages in the dermis (**Figure 3**). Electron microscopy couldn't be done due to lack of facility. The clinico-histopathological features were suggestive of carbon baby syndrome.

Discussion

There are multiple causes of diffuse hyperpigmentation of the skin in infancy. Classifications based on both clinical and histological findings increase the accuracy of diagnosis.[1,2]

Ruiz-Maldonado described a single case of progressive hyperpigmentation in which a child developed pigmentation at the age of three months and became jet black by the age of 4 years. He described him as "carbon baby". Histological examination of this patient showed heavy melanin deposition throughout the epidermis with minimal dermal pigmentation. There was no increase in the

number of melanocytes.[3] Our patients showed similar clinical and histopathological features.

Kaviarasan *et al*.[9] reported a similar case in a 3-year-old Indian girl who developed progressive diffuse hyperpigmentation by the age of 5 months. Histopathology revealed increased melanin deposition in epidermal basal layer.

Furuya and Mishima[4] reported a Japanese child with progressive pigmentary disorder. This child developed hyperpigmentation at the age of 3 months. At 4 years of age the child was mentally retarded with partial hyperpigmentation of the body. Biopsy revealed hyperkeratosis, papillomatosis and proliferation of melanocytes.

Familial progressive hyperpigmentation has been described in kindred. This condition is characterized by hyperpigmented patches that are present since birth and increase in size and number as the infant grows. Most of the skin and mucous membrane surface show increased pigmentation. Microscopically, the melanin granules are more numerous and larger than normal.[5] Familial progressive hyperpigmentation was excluded in our patient as the skin lesions were not seen at birth.

Kint *et al*.[6] described two cases of congenital diffuse melanosis in which the patients developed hyperpigmentation shortly after birth which invaded progressively the trunk and limbs. The pigmentation was diffuse on the abdomen but reticulated on the neck and groin. On electron microscopy, they found the melanosomes were not grouped within the keratinocytes but dispersed within the cytoplasm of the epidermal cells.[6]

Bronze baby syndrome is a rare acquired generalized pigmentary disorder which occurs in the neonates. It is characterized by gray-brown discoloration and occurs in patients with hepatocellular dysfunction undergoing phototherapy. Porphyrin compound undergoes photo destruction which results in a brown substance that is deposited in the skin.[7]

Adrenoleukodystrophy, an X-linked acquired neurodegenerative disease characterised by generalized hyperpigmentation with a slowly progressive involvement of the brain and adrenals. Patient develops uniform macular hyperpigmentation which spares the palms and groin area. This disease is characterized by the accumulation of unsaturated fatty acids with a chain of 24-30 carbons, particularly hexacosanoate in the adrenal cortex and in certain sphingolipids of the brain.[8]

Our patient did not have any evidence of inflammatory condition. We also excluded systemic conditions like Addison's disease, heavy metal toxicity and hemochromatosis from the history, clinical features and laboratory findings.

The etiology of pigmentation in our case is yet unknown. The following hypothesis can be proposed for its occurrence. The role of excessive production of β melanocyte-stimulating hormone, abnormal sensitivity of melanocytes towards normal or abnormal endocrine or neural stimuli and finally to a genetic mutation, which is not yet detected at the chromosomal level. Affection of the two siblings of a family also points towards some genetic factor that might have some role in pathogenesis of this disorder.

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