

Case Report

A NEONATE WITH WHITE FORELOCK AND UNILATERAL RENAL AGENESIS

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ABSTRACT

We present the images of a neonate with white forelock and multiple hypopigmentation at birth. We discuss the differential diagnosis and report a newer association, unilateral renal agenesis.

Keywords: Congenital Piebaldism, Renal Agenesis, Neonate, White Forelock

CASES

A term female baby born to a 23 year old primi, had white forelock and multiple depigmented patches over the central forehead (Figure 1), right flank, anterior aspect of both knees and legs, at birth (Figure 2). Ultrasonogram abdomen revealed absent right kidney. Echocardiogram, bilateral Oto acoustic emission testing and BERA for hearing testing were within normal limits. No other family member had similar lesions.

On follow up, hyperpigmented macules appeared within the lesion (Figure 2) and neurodevelopment was normal.



Figure 1: Neonate with White Forelock (Arrow Heads) and Hypopigmentation Forehead

DISCUSSION

Congenital Piebaldism is a rare autosomal dominant disorder, due to mutations in the *c-kit* proto-oncogene affecting melanocyte development and migration. Clinically, it is characterised by congenital poliosis (white forelock) and well circumscribed chalky white patches, over the central forehead, anterior part of trunk and limbs.

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The dorsal midline, hands, feet, periorificial areas are spared. In due course, hyperpigmented macules appears on both depigmented and unaffected adjacent skin (Janiua *et al.*, 2007).



Figure 2: Same Neonate Showing Multiple Hypomelanotic Patches in Mid Forehead, Right Flank, Bilateral Anterior Knees and Legs at Birth (Arrows)



Figure 3 (Right): Follow up at 3 Months, Hypomelanotic Patches (Arrow Heads) and New Hyperpigmented Macules within the Lesion (Arrows)

Waardenberg syndrome, another autosomal dominant disorder characterised by white forelock, hypertelorism, sensorineural deafness and rarely with Hirschprung disease is a close differential diagnosis (Waardenburg, 1951). Wolf syndrome is a autosomal recessive disorder characterised by Piebaldism and Deafness (Woolf, 1965).

Ziprkowski-Margolis syndrome is a x linked recessive disorder characterised by Deaf mutism, Piebald like lesions over the trunk and extremities and Heterochromia iridis (Ziprkowski *et al.*, 1962).

The presentation at birth, white forelock, multiple anterior lesions with the characteristic hyperpigmented macules within the lesion, normal hearing and static course on followup, differentiate our case (congenital Piebaldism) from other differential diagnosis. Unilateral right renal agenesis in our case is a newer association. Absent family history suggests a denovo mutation.

Clinical Course and Management

Piebald lesions are generally static but contraction of affected lesions and appearance of pigmented macules as in our case can occur. Rarely it is associated with Hirschprung disease, congenital dyserythropoietic anemia type II, Diamond-Blackfan anemia, neurofibromatosis type 1 and Grover disease.

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Medical or light therapy is generally unresponsive in the depigmented skin of congenital Piebaldism. Epidermal cells can be transplanted from donor sites, (mini grafting) can be done. YAG laser therapy can be used for deepithelialization of recipient site. Then, graft can be done over the site (Van geel *et al.*, 2010). Also Autologous cultured epidermal cells can be used for grafting and reported to have 75% success rate with minimal scar (Guerra *et al.*, 2004).

Key Messages

- ❖ White forelock and characteristic distribution of depigmented areas over central forehead, anterior trunk and limbs suggest congenital piebaldism.
- ❖ Absence of abnormal facial features, Ocular, Auditory and Neurological abnormalities differentiate it from other diagnosis.
- ❖ Other systems to be screened for associated anomalies (like renal agenesis in our case).
- ❖ Epidermal cell grafting over affected sites is the effective treatment currently available.

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