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# AYURVEDIC MANAGEMENT OF ERYTHROKERATODERMA VARIABILIS - A CASE REPORT

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#### **ABSTRACT**

Erythrokeratoderma variabilis (EKV) is a rare condition involving keratinization and characterized by well demarcated, pruritic and migratory plaques of erythema and hyperkeratosis. Skin disorders respond better to ayurvedic treatment, but in pediatric age groups the treatment is challenging for several reasons including tendency of the hereditary skin orders to appear anytime from before birth to very young pediatric age group and contraindication of shodhana in children. A 5½ year-old male child was presented with the complaint of erythematous lesions, itching, hyperkeratosis and fine branny scaling on body (symmetrical) since the age of 1 week. During his hospital stay he got complete relief from the erythema, hyperkeratosis and very good relief in distressing itching, making his life easy and comfortable. His improvement was noted during, and after the treatment including follow-up period.

# INTRODUCTION

Erythrokeratodermia variabilis (EKV) are a rare heterogeneous group of inherited cornification disorders. They are characterized by two distinct morphological types of skin lesions; fixed hyperkeratotic plaques and sharply marginated, migratory erythematous lesions. EKV is inherited as an autosomal dominant disorder, rarely autosomal recessive pattern and spontaneous mutation have been reported. Clinically, they manifest as migratory erythema, annular, or hyperkeratotic plaque<sup>1</sup>. Diagnosis of EKV is based on clinical features and genetic analysis. Histopathology findings are nonspecific and include acanthosis, hyperkeratosis,

papillomatosis, and normal granular layer<sup>2,3</sup>. There is no permanent cure for this disease with conventional treatment; only temporary relief can be provided with it. Disfiguring appearance due to hyperkeratosis may lead to psychosocial consequences and psychological complications so this case is reported here as ayurvedic treatment is successful for providing permanent relief in this distressing feature (till the time of submission of this article i.e. for more than 7 months).

### **Case Presentation**

A 5½ year-old child was presented to our hospital with the chronic complaints of erythematous lesions, hyperkeratinization, scaling, itching and skin stretching. They were distributed on the face, chest, neck, trunk, genitals and all the four extremities. Peri-orbital skin stretching led to difficulty in opening the mouth and eating properly. It also leads to restricted movements of facial muscles (due to stretching) and difficulty in facial expression appropriate to situation e.g. smiling or laughing.

His troubles started at the age of 1 week; initially with the erythematous lesions on flexor folds of all the extremities. This gradually increased over the years with involvement of other areas of the body and development of hyperkeratinization associated with itching and scaling. Also erythematous lesions were migratory in nature and in various shapes and sizes that vary with time. The child failed to respond to conventional treatment. The records of previous treatment were not available.

He was born to non consanguineous parents and his natal history was FTND with no history of NICU stay. Family history for similar skin lesions was negative. His developmental milestones were normal. General physical examination was also normal.

On skin examination the erythematous lesions were symmetrically distributed on both sides of face, chest, neck, trunk and on all the four extremities (more intense on flexor aspects), mild itching, skin stretching and hyperkeratinization that were chronic. Cutaneous examination showed well-defined erythematous lesions, hyperkeratotic plaques with diffuse fine scaling distributed on neck, trunk and inguinal fossa (Figure 1a and b). Palms and soles were not involved. Nails, hair and teeth were normal. Systemic examination did not reveal any other abnormality. The patient's IQ and physical growth were normal. Social behavior and academic performance were good.

After complete clinical evaluation the child was put on *ayurvedic* treatment that included *abhayanga*, *swedana* and oral medicaments as per the treatment plan given later. He was daily monitored for his clinical condition and all the observation, irrespective whether improvement or worsening of the features, were recorded.

## Management, outcome and follow-ups

The child was admitted here in IPD of Kaumarbhritya Department. Orally he was given kanchanar guggulu<sup>4</sup> throughout his stay in the hospital. Abhyanga with jatyadi taila<sup>5</sup> was daily done as initially there was much scaling, and in the later part of his stay to prevent scaling in the areas of hyperkeratinization. After about two week-periods when scaling was almost cured he was advised to apply bala taila<sup>6</sup> to get rid of hyperkeratosis. This was in addition to abhyang with jatyadi taila. The treatment duration in the hospital was of one month.

During this period a couple of small blisters appeared on the abdomen, but they healed within two days with local dusting of powder of triphala, nimba twak and haridra<sup>8</sup>. Subsequently no new blisters developed. On 8<sup>th</sup> day the child developed confluent furuncles on the scalp that also healed after two days with the same dusting. The child became completely free from hyperkeratosis and scaling (from 22<sup>nd</sup> day of treatment in the hospital) and continues to remain free from those complaints for more than four months after discharge from the hospital (as observed during three consecutive follow-ups) (Figures 2, 3, 4 and 5). Itching was only negligible (sometimes present; sometimes absent) while erythema, which was completely relieved earlier (on 22<sup>nd</sup> day), but appeared again at two different locations (around anus and near the penile base at two different times. Anal erythema cleared within less than a week time while penile erythema cleared after discharge from the hospital and the child remained free from it in the summer months. However, it recurred at different areas (in the folds of all the extremities) during the beginning of monsoon season.

# Treatment plan

# **Oral medication:**

1. Kanchanar guggulu 1 tablets x bid with water after food

Topical medication:

- 2. Abhyanga with jatyadi taila followed by bashpa swedana
- 3. Local application of *bala taila* after scaling and erythema were cleared (two to three times) daily

- (from 16<sup>th</sup> day of starting treatment to be very precise) to clear hyperkeratinization
- 4. Dusting with powder of *triphala*, *nimba twak* and *haridra* (all mixed in equal amount by weight) SOS (used only for blisters and furuncles)

# **DISCUSSION**

At present there is no curative treatment; only palliative treatment is available for moisturizing the affected part of the skin and antihistaminics, if there is itching. Genetic analysis could not be performed for the obvious reasons of heavy financial implications. Skin biopsy was also not done as it is not considered to be a diagnostic criterion as there are no distinctive features. Clinical diagnosis is of much importance and so the diagnosis was done exclusively based on clinical features.

Congenital skin disorders usually run a long course and the disfiguring appearance may lead to psychosocial consequences and psychological complications so it is felt that the case deserves publication or presentation for discussion. With minimal oral medicines and abhyanga along with some topical application of appropriate medicated ghrita or taila the disease can be managed well. Bashpa swedana would be helpful in making the skin soft. Scaling responded well to abhyanga with jatyadi taila. Remarkably, hyperkeratinization got under control very well with addition of topical application of bala taila and till last observation (at the end of four months after discharge) it remains under control. Erythema does keep appearing and disappearing and true to its character it remains migratory. However, the area that it covered was far more less than it covered before treatment. Also it is felt that the erythema aggravated due to humid and hot climate as the areas where there would be more sweating were involved in erythema. Simple medication (dusting) is also remarkably effective in controlling blisters and furuncles. They completely healed within two days. Bala taila (just local application) was used for clearing hyperkeratosis keeping in mind that Sushrut has mentioned its use in neonates for abhyanga. Also the cases of ichthyosis (number less than 10) under this kind of treatment in our hospital have responded very well. Hence it was thought

that local application of *bala taila* may keep the patient's skin smooth and the logical application of this knowledge worked.

# CONCLUSION

Skin disorders respond better to *ayurvedic* treatment, but in pediatric age groups the treatment is challenging for several reasons including tendency of the hereditary skin orders to appear anytime from before birth to very young pediatric age group and contraindication of *shodhana* in children. Contemporary treatment could not provide satisfactory results. Patient got complete relief from the erythema, hyperkeratosis and very good relief in distressing itching, making his life easy and comfortable *Ayurvedic* treatment was sought to get relief from this highly distressing skin condition.

**Informed Consent:** Informed consent was taken from the father since the patient is a minor.

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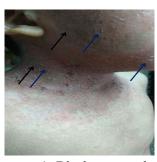




Figure 1 A and B (Before treatment): Black arrows show hyperatinization and blue arrows show scaling on face, chest, neck, trunk, genitals and all the four extremities





Figure 2 A and B (After treatment): Note the clearance of keratinization on the face and ears







Figure 3 A, B and C (After treatment): Note the clearance of keratinization on trunk and four extremities

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