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A REPORT ON XERODERMA PIGMENTOSUM IN KARNATAKA

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ABSTRACT

Four suspected cases of a rare disease, Xeroderma Pigmentosum, have come to light in Kollegal Taluk of Karnataka in 2016. It is characterized by skin pigmentation, lesions on face including the head and inability to see in bright light, leading to blindness. This brings out the importance of skin as the natural protective layer of the body. This article is a report on this rare disease.

Keywords: Xeroderma Pigmentosum, *Kushta*, Lesions, Blindness, Skin Cancer.

INTRODUCTION

Prajavani, a Kannada daily reported in 2016 that a special type of skin disease was noticed among children in *Kollegal* Taluk of Karnataka state. It was reported that for the last fifteen years this unique skin disease was not treated properly leading to the death of the affected children in their teens. This prompted a field visit to *Kuratti- Hosur* village (7 km from *Kowdalli* on the road to *Malai Mahadeshwara* Hills from *Kollegal*) on 21-07-2016 by a team consisting of Dr. Sathyanarayana Bhat, Dr. Pratibha, V. V. Bhat and Venkatachalapathi. Sri Venkatarama Shetty who facilitated our visit informed us that four patients of this skin disease have expired before they crossed the teens. There were only four living patients. The following information was collected in discussion with the parents of the affected children; after visiting their houses on 21-07-2016:

1. Girl aged 6 years, Parents married in close circle. First male child is healthy. Second female child developed pigmentation of skin in 9 months after birth. Symptoms: Black spots on the skin all over the body; Redness in eyes with limited visibility, Inability to see light (photophobia), Bent neck. Normal in activity,

Shy. Treated for two weeks in Govt. College of Ayurveda Hospital, Mysuru.

2. Girl aged 16 years, Bullosa in head and wound/ lesion in nose. Severe since last 5 years, Spots on the skin all over the body. Totally blinded. Loss of hair on head. Weak constitution. Not a case of marriage in close circle.
3. Boy aged 3 years, unable to see light, abnormal skin features on the face (He was not present in the village during this visit. However, he was subsequently treated for two weeks in Govt. College of Ayurveda Hospital, Bengaluru).
4. Boy aged 2 years was not seen. Three children of this family had expired, the last one in June 2016, after a belated visit to Ayurveda Medical College Hospital- Mysuru. Not a case of marriage in close circle.

The photographs of the above three cases are given in annexure –I

Symptoms

All the cases had the following symptoms:

a) Discomfort and inability to see light in bright light, leading gradually to blindness.
b) Spots in the skin.
c) Lesions, predominantly all over the face including the head.

Diagnosis

Based on these features and reference to the pictures available in the Google Images, it was initially felt that this rare disease may be a variety of Epidermoloysis Bullosa. On discussion with the Dr. S Narahari, Dermatologist, he suggested that it could be Xeroderma Pigmentosum. After reference to the Google Images, we felt that the symptoms of these cases fit into the description of Xeroderma Pigmentosum.

Xeroderma Pigmentosum is characterized as an autosomal recessive disorder and caused by abnormality in genes. It is reported that Ultraviolet light such as from sunlight damages the genetic material (DNA) in skin cells. This damage leads to patches on the skin (pigmentation) and in due course leads to skin cancer. Inability to see in light (Photophobia) and pigmentation usually occurs before the child is 5 years old and the life span is limited to about 15 years. Except total protection from sun light (with dark glasses and clothing) no other effective medicine is reported.

AYURVEDIC PERSPECTIVE

The non availability of any known remedy for the above rare disease made us to revisit the Ayurvedic literature about skin disease. The skin is the natural protective layer of the human body, which also functions as a sensory organ, excretes sweat, and helps in body temperature regulation.

The seven layers of the skin *'twak'* are formed in layers like in boiling milk' *"Tasya khalvevam pravrittasya kshurasyeva santavikaah sapta twacho bhavanti."* (4/3- *Sushruta Samhita*).These are listed as *Avabhasinee, Lohita twak, Sweta twak, Taamra twak, Vedinee, Rohinee* and *Mamsadhara*. However, *Charaka Samhita* has listed only six layers with different names.

Skin diseases are mentioned in Atharva Veda. *Kushta nashana Sukta* (1-23) mentions three types of skin disease called '*Kilasa*', '*Palita*' and '*Nirita*' which can be cured with 3 types of Curcuma longa- (*Rama Rajani, Krishna Rajani and Asikni Rajani*). *Shweta Kushta nashana Sukta* (1-24) mentions cure of Leprosy through use of *Asuri, Sarupa* and *Shyama* plants. Among other reasons, *"Purakrita karma"*(actions which are previous and acquired) and *"Papakarma"*(undesirable and wrong actions) are also considered as causes of skin diseases, particularly those considered "*asadhya*" or difficult/ impossible to be cured.

Charaka Samhita lists 11 *Mahakushta* and 7 *Kshudra kushta* varieties. Text books of Ayurveda have listed the following varieties, which are difficult to be clearly diagnosed in practical terms. Therefore it is difficult to provide the possible English identifications of those conditions.

***Mahakushta* (Leprosy)**

1. *Kapala.* (Krishna arunam kapalabham- black and reddish skin, like broken pieces of earthen pot, Dry, rough and thick skin lesions with excessive pain)
2. *Mandala* (Ring worm).
3. *Audumbara.* (Burning sensation, itching, pain, redness brown hair on the patches resembling fruit of Audumbara.)
4. *Rishya jihva.* (Rough red edges, brown inside, painful, resembling the tongue of antelope (Rushya.)
5. *Pundarika.*(White in color with red edges resembling the leaf of lotus and elevated with burning sensation.)
6. *Sidhma.* (discharging powder upon touching it, generally located in chest area, resembling the flower of bitter gourd (alābu) white and coppery in color)
7. *Kakanaka* (Red in color resembling the seed of Gunja, extremely painful)

Kshudra Kushta

8. *Eka kushta* (Psoriasis) (Absence of sweating, extensive localization, resembling the scales of fish)
9. *Charmaakhya* (Skin over the patch becoming thick like the skin of the elephant)
10. *Kitibha* (Psoriasis?) (Blackish brown in color, hard and rough in touch like scar tissue)
11. *Vaipadika* (Rhagades) (Cracks in palms and soles of feet with excruciating pain)
12. *Alasaka* (Lichen) (nodular growth, with excessive itching sensation and redness)
13. *Dadru* (Circular patches with elevated edges, itching sensation, redness and pimples*)*
14. *Charmadala*. (Redness, itching, boils, pain, cracks in the skin, tenderness and painful)
15. *Pama* (Eczema Dermatitis) (Excessive itching, eruptions which are white, reddish or blackish brown in color)
16. *Visphota.*(Exanthymata Bullosa) Viral Impetigo (white, reddish boils, thin skin over rash)
17. *Shataru* (Rupia). (Red, brown, burning sensation, painful, many lesions)
18. *Vicharchika* (Pemphigus) (Bacterial) (blackish brown eruptions, itching sensation, excessive exudation)
19. *Kilasa (Sweta Kushta*)- Vitiligo (Leucoderma) (depigmentation of skin leading to white patches, more particularly in hands, the neck, face, back and wrists)

DISCUSSION

Since 2008 Rare Diseases Day is being observed annually on the last Day of February. The awareness about non-availability of sufficient or proven treatment for rare diseases and research into such diseases are inadequate. Therefore the observance of Rare Diseases Day is expected to generate scientific curiosity and spirit of research. Perhaps patient advocacy and use of internet and blogging have helped the families of sufferers of rare diseases to share information and experience. Information- mining from the internet shows considerable amount of experience sharing by the affected people and their families. No longer can we classify some of them as incurable syndromes or *"asadhya"*. What can be more tragic than a medical researcher not taking up a disease for research because of prima facie understanding that a problem is considered as *"asadhya"*. by the previous generation?"Sadhyasadhya vibhaga" mentioned in Ayurvedic literature is only a general advice to the practitioners. It should not be a constraint for the scholars and researchers. No progress would have taken place in the art and science of medicine if researchers had not taken up investigation of diseases considered as not curable. Therefore, we feel that how so ever challenging research into a rare disease like this, it is scientifically important to undertake such research efforts.

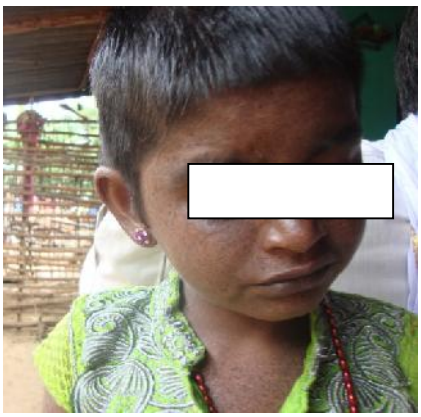
CONCLUSION

Ayurvedic literature and clinical practices do not enable us to categorically identify the different kinds of skin diseases covered under general expression *Kushta* (Leprosy). Certain types of skin

diseases are curable while some others are not. The difficulty involved in identification/ diagnosis of the types of skin diseases makes it difficult to identify the possible remedy described in Ayurveda. We hope that practitioners and researchers of medicine may take up further study and research into this rare disease.

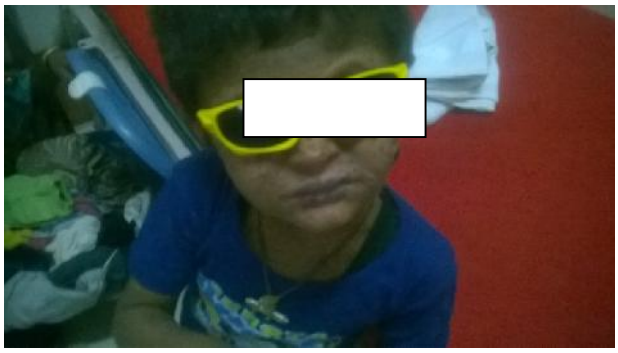
Cases of Xeroderma Pigmentosum

Case 1



Case 2

Case 3



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