



Review Article

WEREWOLF SYNDROME –An Orphan Genetic Disorder

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Hypertrichosis is expressed as a genetic disorder in which hair follicles that react to androgens. This will allow for the heaviness of the hairs and rate of growth to increase. There are many other different causes for a person to develop this orphan disorder like congenital hypertrichosis and acquired hypertrichosis. The overall cause of hypertrichosis for the most part is unknown. Scientists consider that hypertrichosis is a form of genetic disorder that arise due to a spontaneous mutation in the DNA sequence. However, some studies have shown that the mutation leading to congenital generalized hypertrichosis (CGH) is located between q24-q27 regions of the X-chromosome. There are many affordable and convenient treatment options for hypertrichosis. They include shaving, trimming, plucking, bleaching, waxing, chemical depilatories, electrosurgical epilation, and topical eflornithine. Recently, laser hair removal has been proposed as a treatment option. It is believed to have less side effects and produces long-lasting results.

Key words: Hypertrichosis, Werewolf syndrome, genetic disorder.

1. INTRODUCTION

Hypertrichosis is the condition used for the enhancement of hair on any part of the body in excess. Hypertrichosis may either be an isolated finding or be associated with other abnormalities¹. Hence, appropriate diagnosis of hypertrichosis into definite classification is essential. Excessive hair may cause beauty concern, cosmetic unease, resulting in a serious emotional stress load, importantly if extensive. Treatment options are finite, and the results of treatment may not be always satisfactory. Therefore, patients should be appropriately advised of the available treatment options. Present available treatment options include cosmetic procedures like bleaching, trimming, shaving, plucking, waxing, chemical depilatories, and electrosurgical epilation and laser. Hair removal through laser is the most promising method for long-term hair removal. In general, treatment of

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2. EPIDEMIOLOGY

Congenital hypertrichosis is rare as compared to acquired modes of hypertrichosis. Only 50 cases of congenital hypertrichosis have been recorded since the middle Ages, and fewer than 100 cases of congenital generalized hypertrichosis have been documented in scientific publications². Acquired hypertrichosis and hirsutism are more common and occurs in about 10% of women between ages 18 to 45. However, a large number of cases of acquired hypertrichosis are drug induced (especially due to minoxidil) have been diagnosed³.

HISTORY

The foremost case of hypertrichosis was observed in Petrus Gonsalvus of the Canary Islands which was documented by Altrovandus in 1648. He noted that two daughters, a son, and a grandchild in Gonzales' family all had hypertrichosis². During the next 300 years, about 50 cases were observed. The scientist Rudolf Virchow illustrated a form of hypertrichosis accompanied by gingival hyperplasia⁴.

3. CLASSIFICATION

I. Congenital Hypertrichosis⁵

a. Congenital hypertrichosis lanuginose:

This is a very rare type with only about 50 cases reported worldwide since the middle Ages. This condition is portrayed by extravagant hair growth by birth itself. More proportion of the body is topped with lanugo hair, which is a fine, soft, unpigmented and silky hair that coats the fetus and which is usually shed at around eight months of gestation and replaced with fine vellus hair and terminal scalp hair in preparation for birth. In congenital hypertrichosis, lanugo hair progress to grow and this exorbitant long fine hair present throughout the life.



Fig 1: Congenital hypertrichosis lanuginosa

Congenital hypertrichosis terminalis:

This is a alteration of congenital hypertrichosis which ally with hair growth all over the body, but the hair is fully pigmented and the condition is accompanied with gingival hyperplasia⁴



Fig 2: Congenital hypertrichosis terminalis
Naevoid hypertrichosis:

This is an uncommon form of hypertrichosis where circumscribed area of terminal hair growth occurs. It is not usually accompanied with any other disease, except if it emerges as a faun-tail on the lower back, when it may indicate underlying spina bifida. Naevoid hypertrichosis can arise at birth or appear later stages of life. Example of naevoid circumscribed hypertrichosis is the presence of a solitary and bushy eyebrow.



Fig 3: Naevoid hypertrichosis
Hypertrichosis associated with naevi:

Hypertrichosis is a characteristic aspect of congenital melanocytic naevi, vascular malformation, Becker naevi and less frequently, other birthmarks.



Fig 4: Naevoid hypertrichosis and Melanocytic naevus

II. Acquired Hypertrichosis

Acquired hypertrichosis characterize excess hair growth which evolves in an individual after birth. Mostly the hair is unpigmented or may involve pigmented terminal hair. Excess hair growth may be generalized or localized which covers all hair-bearing areas of the body.



Fig 5: Drug-induced Hypertrichosis

ETIOLOGY⁷⁻⁹



SIGNS AND SYMPTOMS

The hallmark feature of hypertrichosis is surplus hair on the various parts of the body. Hair in hypertrichosis is unusual and not normal in length, density and may consist of various hair types such as lanugo, vellus or terminal hair¹. Patterned forms of hypertrichosis cause hair growth in patterns². Generalized forms of hypertrichosis shows hair growth all over the body. Whereas, localized forms lead to hair growth restricted to a certain area.

4. PATHOPHYSIOLOGY

Various mechanisms can lead to hypertrichosis. One among them affects areas of the skin that are switching from the small vellus type to the larger terminal type². This process usually happens during adolescence, when vellus hair follicles in the underarms and groin grow into terminal hair follicles¹. Hypertrichosis involves this same type of switching, but in areas that do not normally produce terminal hair. The mechanisms for this switch are poorly understood. Another mechanism involves a change in the hair cycle. There are three phases of hair cycle i.e., hair growth phase, hair follicle death phase and the hair shedding phase. If the hair growth phase increases beyond what is normal; that region of the body will experience extravagant hair growth¹⁰.

DIAGNOSIS

Diagnosis is completely based on the observation of clinical presentations of excessive hair on the body parts. No specific investigation is required¹¹.

5. MANAGEMENT OF WEREWOLF SYNDROME

Treatment for hypertrichosis is basically hair removal. Several methods are available but need to be repeated regularly as hair continues to grow back which can also cause dermatitis, scarring or hypersensitivity reactions as well⁹. The major treatment for hypertrichosis involves cosmetic procedures which basically remove the excessive hair from the body through various methods such as

plucking, bleaching, trimming, shaving, waxing, chemical depilatories, electrosurgical epilation, and laser therapy. No single method can be chosen as a best fit for removing the hair, because it depends on the hair type, the amount, area, and the patient themselves¹¹.

I. NONPHARMACOLOGICAL MANAGEMENT
Chemical Depilatories:¹²

It is a method that breaks the hair at the surface of the skin. This method acts by affixing to the hair, abolishing the oily layer on the shaft, and making the thioglycolates to break down the keratin in the hairs. This method is usually used weekly on small areas.

Electrosurgical Epilation:

It is permanent method of removing the hair on the body parts. Usually this procedure is done by inserting a fine needle into the hair follicle and electrical currents are sent through it to destroy the dermal papilla or follicle of the hair. There are two types of options that are used in this method i.e., electrolysis and thermolysis. Electrolysis wipeout the dermal papillae layer of the follicle through electrical currents. Electrosurgical epilation will prevent most hairs from growing back.

Laser Treatment:¹³

This is typically used for long-term effects. In this method, high amount of pulse light is consumed by the malanosomes of the hair bulb. This light energy is then emanating the bulb as heat damaging the surrounding tissues causing the hair not to re-grow. This is less painful than electrosurgical epilation and usually more effective.

PHARMACOLOGICAL MANAGEMENT

Tropical Eflornithine:¹⁴

An innovative recent treatment option available for slowing down the excessive hair growth is topical eflornithine, an inhibitor of ornithine decarboxylase enzyme present in hair follicles which plays a vital role in hair growth process. This treatment is applied as a cream. This is generally applied over the affected area after laser therapy to make it more effective.

There is no permanent cure for hypertrichosis. Therefore, people with hypertrichosis must undergo endless treatments to remove the excess hair throughout their lifetime¹⁵.

6. DISSCUSSION

An orphan disease is defined as a condition that affects fewer people worldwide. This includes diseases as familiar as Tired All The Time Syndrome (Chronic Fatigue Syndrome), Beauty Parlour Syndrome, Fat Wallet Syndrome, Male Breast Cancer, Selfie syndrome and many more are of increased concern and clinicians must be made aware of these Orphan diseases for better diagnosis and management¹⁶⁻²⁰.

There is a deficit of medical and scientific knowledge related to these diseases. Doctors, researchers and policy makers were unaware of most of the rare diseases and until very

recently there was no real research concerning issues related to the field. There is no permanent cure for most of the orphan diseases, but the appropriate treatment and medical care can improve the quality of life of those affected with them.

7. CONCLUSION

Although this clinical condition is uncommon, it can lead to various conditions if undiagnosed and untreated. In particular, the severe consequences of these disorders necessitate a major effort to better define the clinical focus on the pathogenesis and to develop pharmacological treatments. Hence, if there is awareness among patients and clinicians related to this disease will definitely help to improve the current status of medical care.

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