

**WEREWOLF'S SYNDROME – CASE REVIEW****Deepika R^{*}, Iram Naz Ansari, Mohammed Bilal, Amina Jabin, Kesiya Simon, Anjana Sankar and Zachariah Thomas**

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ABSTRACT

Werewolf 's syndrome, which is scientifically known as hypertrichosis is an unusual genetic disorder. It is characterised by abnormal uncontrolled hair growth, which can be either generalized (all over the body) or localized (confined to certain parts). WWS is classified mainly as congenital or acquired. Congenital hypertrichosis is a condition, where babies are born with abnormal uncontrolled hair growth. In acquired hypertrichosis, hair growth develops later in life due to lifestyle or some drugs. Some types of congenital hypertrichosis are hypertrichosis lanuginosa, generalized hypertrichosis, terminal hypertrichosis, localized hypertrichosis, nevoid hypertrichosis and circumscribed hypertrichosis. Acquired hypertrichosis is classified as acquired hypertrichosis lanuginosa, acquired generalized hypertrichosis, acquired patterned hypertrichosis, and acquired localized hypertrichosis. Common symptoms are abnormal hair growth and teeth defects, enlarged gums (Gingival Hyperplasia), mental retardation etc. Most of the hypertrichosis are caused due to genetic reasons. Acquired werewolf syndrome also can be caused by cancer or as side effects of certain medications (latanoprost, cyclosporine, acetazolamide, streptomycin). Congenital hypertrichosis is rare, as compared to the acquired ones. Few types of werewolf syndrome can only be controlled with treatment; while others are managed with cosmetic hair removal, depilatory creams, and electrolysis. Further research is required to establish an effective treatment regimen to patients with WWS.

Key Words:- Werewolf's syndrome (WWS), Hypertrichosis, Gingival Hyperplasia, Hypertrichosis Lanuginosa, Nevoid Hypertrichosis, Circumscribed hypertrichosis, Acquired Patterned Hypertrichosis.

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Email:- deepikardileep@gmail.com**INTRODUCTION**

Hypertrichosis, also known as werewolf syndrome, is a condition characterized by abnormal hair growth anywhere on a person's body. It can affect both women and men. The abnormal hair growth may cover

the face and body. Hypertrichosis is informally been called werewolf syndrome because the appearance is similar to that of werewolf (Wendelin D *et al.*, 2003). It is also associated with additional anomalies such as gingival hyperplasia, deafness, cardiomegaly and bone abnormalities. Hypertrichosis is different from hirsutism. In hypertrichosis, there is only little or no new hair growth but in hirsutism, new hair growth is continually growing (Krishna S, 2017). Some types of acquired werewolf syndrome can be caused by cancer or as side effects of certain medications (latanoprost, cyclosporine, minoxidil, acetazolamide, streptomycin) (Wysocki G and Daley T, 1987; Demitsu T *et al.*, 2001; Burton J and Marshall A, 1979; Chellini P *et al.*, 2015). WWS is classified as congenital or acquired. The currently available treatment methods include cosmetic procedures (bleaching, trimming, shaving, plucking, waxing, chemical depilatories, and electrosurgical epilation), and hair removal using light sources and lasers. Laser-assisted hair removal is the most efficient

method of long-term hair removal currently available (Alajlan A *et al.*, 2005).

EPIDEMIOLOGY

In most hypertrichosis cases, men and women are equally affected. Congenital hypertrichosis is rare compared to acquired ones. However, many cases of acquired hypertrichosis are drug induced (especially due to minoxidil, latanoprost) have been diagnosed (Krishna S, 2017).

TYPES OF WEREWOLF SYNDROME

Werewolf syndrome is scientifically known as hypertrichosis. It is classified on different basis. They are as follows:

- Extent of distribution
- Age of onset
- Site involved

Types of hypertrichosis

Based on extend of distribution

- A. Generalized: When excessive hair growth covers your entire body.
- B. Localized: When the excessive hair growth is restricted to a particular area.

Based on age of onset

1. Congenital hypertrichosis
2. Acquired hypertrichosis

Congenital hypertrichosis is the condition, where babies are born with abnormal hair growth. In acquired types, such hair growth develops later.

CONGENITAL TYPES: Congenital hypertrichosis is categorized as follows

- Hypertrichosislanuginosa
- Generalized hypertrichosis
- Terminal hypertrichosis
- Localized hypertrichosis
- Nevoid hypertrichosis
- Circumscribed hypertrichosis

Hypertrichosis lanuginosa

In congenital hypertrichosislanuginosa, the newborn has lanugo hair all over the body, except palms of hands and soles of feet. Hair thinning also happen.

Generalized hypertrichosis

In this type, excess facial and upper body hair in males, and an asymmetrical hair growth in females is found.

Terminal hypertrichosis

This type is characterised by growth of highly pigmented terminal hair, all over the body.

Localized hypertrichosis

Here hair growth is limited to certain areas only.

Circumscribed hypertrichosis

It is the unusual hair growth of the upper extremities (shoulder, arms, elbow, wrist). Mostly,

elbows are found to be more involved in this type of werewolf syndrome, which is also known as hairy elbow syndrome.

Nevoid hypertrichosis

In this type, it features an excessive hair growth on an isolated area.

Nevoid hypertrichosis has a variable etiology.

1. Primary nevoid hypertrichosis occurs in the absence of extracutaneous associations.
2. Secondary nevoid hypertrichosis is associated with lipodystrophy, hemihypertrophy, scoliosis, and abnormalities of the underlying vasculature (Pavone Pet *et al.*, 2015).

ACQUIRED TYPES

This type of werewolf syndrome is classified as:

- Acquired hypertrichosislanuginosa
- Acquired generalized hypertrichosis
- Acquired patterned hypertrichosis
- Acquired localized hypertrichosis

Acquired hypertrichosislanuginosa

In this case, the affected person experiences rapid lanugo hair growth on different parts, especially the face.

Acquired generalized hypertrichosis

Acquired generalized hypertrichosis refers to the unusual and abnormal hair growth on different parts of the body like, the cheeks, chin, upper lips, forearms, and legs (Wendelin D *et al.*, 2003; Sunil M *et al.*, 2016).

Acquired patterned hypertrichosis

Here the rapid hair growth will be in some specific pattern.

Acquired localized hypertrichosis

Localized hypertrichosis is the abnormal hair growth restricted to certain areas only (Horning G *et al.*, 1985).

Drug Induced Hypertrichosis

Latanoprost used for the treatment of glaucoma has many side effects like darkening of the iris, hypertrichosis and hyperpigmentation of the eyelashes (Demitsu T *et al.*, 2001).

Hypertrichosis is a cosmetically undesirable side-effect of cyclosporine therapy. The cessation of cyclosporine therapy resulted in a progressive resolution of the induced hypertrichosis (Wysocki G and Daley T, 1987).

Minoxidil induced Hypertrichosis

Hypertrichosis is a common side effect of topical minoxidil (Burton J and Marshall A, 1979; Chellini P *et al.*, 2015).

ETIOLOGY

Congenital generalized hypertrichosis is a feature of several inherited syndromes in which genetic errors resulting in dysfunction of proteins that may lead to the development of the hair follicle. Some evidence to

support that exposure to medications such as minoxidil in uterus may predispose to congenital generalized hypertrichosis. Drugs most often cause acquired generalized hypertrichosis.

- Antibiotics such as streptomycin
- Vasodilators (diazoxide, minoxidil, prostaglandin E1)
- Diuretics (acetazolamide)
- Anticonvulsants (phenytoin)
- Immunosuppressives (cyclosporine)
- Chelators (penicillamine)
- Latanoprost

DIAGNOSIS

A detailed review of patient history and an in-depth physical examination are necessary to determine the presence of abnormalities. Particular attention should be given to the presence of other anomalies in particular the face, eyes, teeth, heart, kidneys, bones, and extremities. The diagnosis is directed for identifying the signs associated with Congenital Hypertrichosis and distinguishing it from the acquired form. Depending on the clinical manifestations, laboratory analyses, skeletal X-rays, Brain-MRI, ultrasound EEG, ECG and echocardiogram, and psychometric tests are useful investigations.

TREATMENT

Management and treatment of hypertrichosis

Hypertrichosis is a cause of significant emotional distress for the affected patients and their family. The cosmetic embarrassment is relevant when the hair is widely distributed over the areas of the body normally uncovered. The different approaches are:

- 1) Cosmetic procedures
- 2) Intense pulsed light source and laser treatment
- 3) Pharmacological treatment

Not all the treatments are effective over the long term, so the choice of therapy should be made taking into consideration aspects such as the location of the excess hair, its association with complex anomalies and age of the patients.

Cosmetic procedures

Cosmetic procedures includes using bleaching methods to make dark-colored hair less evident. And different procedures are used to remove the excess hair, such as trimming, shaving, plucking or waxing. Clinical depilation acts by damaging the hair directly on the skin surface. Electrosurgical epilation is more effective (Alajlan A *et al.*, 2005). The use of shaving or clinical hair removal may give excellent results but it may also cause irritation and allergic contact dermatitis.

Intense pulsed light sources

Effective results are obtained by intense pulsed light sources and various type of lasers (Ruby, Alexandrite, Diode). A Neodymium Yttrium-Aluminum Garnet (Nd:YAG) laser has been used in the removal of excess hair (Alajlan A *et al.*, 2005). Good results can be achieved with electrolysis or Alexandrite laser. In some cases, depilation may be definitive.

Pharmacological treatment

It consists of topical eflornithine, a specific and irreversible inhibitor of the enzyme ornithine decarboxylase, which stimulates hair growth. Cosmetic treatment is always useful to avoid the social rejection and isolation of patients. In one third of cases, regrowth of the hair may occur, thus necessitating further procedures (Alajlan A *et al.*, 2005).

Fig 1. Hypertrichosis

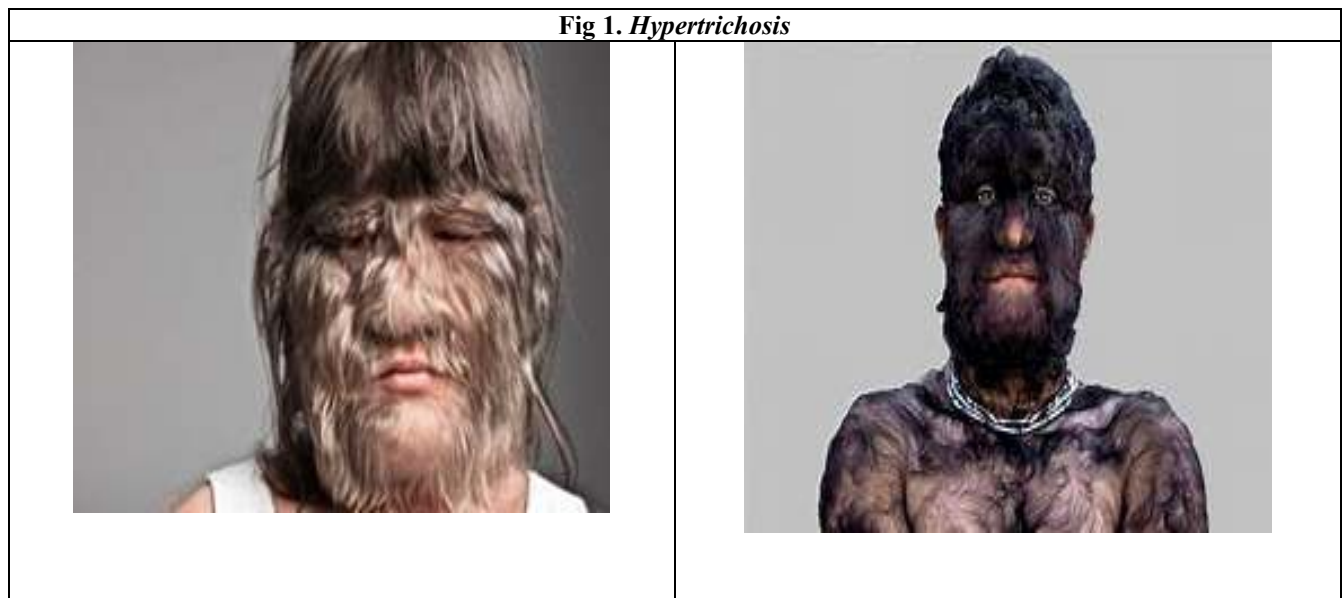


Fig 2. Congenital hypertrichosislanuginose



Fig 3. Generalized hypertrichosis



Fig 4. Terminal hypertrichosis



Fig 5. Localized hypertrichosis



Fig 6. Circumscribed hypertrichosis



Fig 7. Nevoid hypertrichosis



Fig 8. Latanoprost induced Hypertrichosis



Fig 9. Cyclosporine induced Hypertrichosis

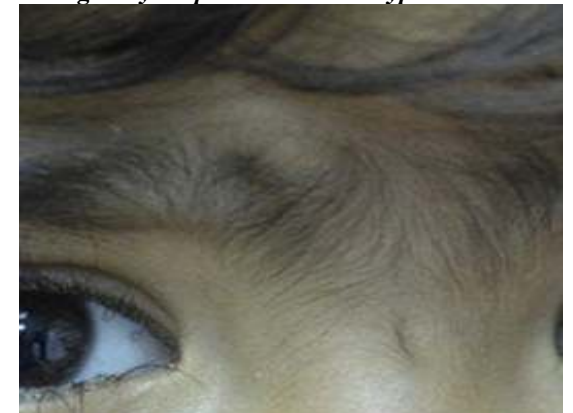


Fig 10. Minoxidil induced Hypertrichosis

DISCUSSION

Hypertrichosis is a rare disorder that causes abnormal excessive growth of body hair. Here excess growth of hair occurs in areas of the skin with the exception of androgen-dependent hair of the pubic area, face, and axillary regions. In contrast, hirsutism is characterized by excessive growth of androgen-dependent hair. Gingival enlargement is one of the main oral manifestations and a common feature. Hypertrichosis is also associated with severe mental retardation, clouding of cornea, enlargement of liver and spleen etc (Anavi Y *et al.*, 2008). Long-term removal of hair is a challenging concern. Epilatory methods of hair removal would be a better method than the depilatory methods because of the longer duration of hair free skin. Newer therapies using lasers and lights is very helpful for removing excessive hair growth. As this disorder is so rare, large studies in this aspect is lacking. Long-term treatments include electrolysis and laser surgery. Electrolysis is destruction of individual hair follicles with small electrical charges. Laser surgery involves the application of a special laser light over several hairs at

one time (Alajlan A *et al.*, 2005).

CONCLUSION

Hypertrichosis is a wide topic in the field of clinical practice. To conclude, hypertrichosis on its own is still an enigma, which needs further research to get a detailed understanding of this context. Hypertrichosis can cause severe emotional distress for patients, especially those who do not have access to permanent laser hair removal or electrolysis. Self-confidence and quality of life will be low for these patients. In cases of severe hypertrichosis, it is important to arrange mental health care for patients in addition to medical care to address the underlying cause of the hypertrichosis. For those with inherited disorders, there is no cure and poor cosmesis is a lifelong issue. For those with acquired hypertrichosis, the outcomes are good. Although this clinical condition is uncommon, it can lead to various conditions if undiagnosed and untreated. Hence, if there is awareness among patients and clinicians related to this disease, it will definitely help to improve the current status of medical care (Sunil M *et al.*, 2016).

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