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Pharmaceutical Sciences **Review Article……!!!**

AN UPDATED OVERVIEW ON AMBRAS SYNDROME

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ABSTRACT

The amount of unpopular, “scientific” werewolf theories is inversely proportional to serious investigations into the history of the werewolf. A critical approach of these theories leaves the scientist, dermatologist and the very common people starving for clear explanation. It was found decades ago but explained 20 years ago. This review will help us to increase and expand our knowledge towards this panic syndrome, and should encourage future dermatologic analysis to report more on Ambras syndrome.

INTRODUCTION:

Hypertrichosis or ambras syndrome is a key solution to the early mysterious myth formally called werewolf syndrome (The person who was covered by the layer of thick coat of fur) because the appearance is similar to the early mythical werewolf. Hypertrichosis can be either congenital (present at birth) or acquired later in life. The 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. It results in the increased growth of vellus or other hair at inappropriate locations. The affected area of the body contains numerous amounts of hair follicles than elsewhere. This condition is unrelated to virulisim or any menstrual abnormalities found in female. The occurrence of hair varies depending upon the patient age groups.

Estimated causes of ambras syndrome

Although there are no sure answers to those questions, many believe that the condition is to be genetic disorder. It can be considered to be inherited or just happen due to mutation. Some believe it occurs after individual have cancer, is also caused by usage drugs or metabolic disorder as well.

Human is not the only prey [1]

This condition is also found in canine species , A male ***persian*** cat with the condition named Atchoum achieved a certain level of notice due to the unusual appearance of hypertrichosis gave him, and has been nicknamed “The Werewolf Cat”.

Famous historical ambras syndromians :

- Fedor Jeftichew ("Jo-Jo the Dog-faced Man")
- Stephan Bibrowski ("Lionel the Lion-faced Man")
- Annie Jones ("the bearded woman")
- Alice Elizabeth Doherty ("The Minnesota Woolly Girl")

Society and culture

The society never allowed anyone to grow as wellbeing, they never encouraged them to show off their real faces in the light, and they lived the life filled with pain and fear.

Works they have assigned

They were considered as a curse and never allowed to enjoy the normal life and worked as jokers in circus, the earned money by performing in shows and as barber in the shops during that period of time.

Evidence from the holy book BIBLE (Hypertrichosis of Esau)

"The first to emerge was reddish and his whole body was like a hairy mantle". So they named him Esau (Hairy). Genesis 25:25. New International Version (NIV). The story of Jacob and Esau is told in the Old Testament book of Genesis (Genesis 25:19-33:20. NIV). Jacob and Esau were the twin sons of Rebekah and Isaac. The story is one of struggle starting before birth and through their adulthood. Esau, we are told, was the first born and was covered with red hair "like a hairy mantle." Other translations are "garment," "fur coat," while others translate it as "it (hair) was all over his body."

The first observed case

After the case observed in the bible, it reappeared in the period of 1648. His name was mentioned as "Pertrus Gonsalvus" (figure no 1) of the Canary Islands. This was documented by Altrovandus in mid-17th century and published in his text Monstrorum Historia cum Paralipomenis historiae omnium animalium published in 1642.] He noted two daughters, a son, and a grandchild in Gonsalvus' family all had hypertrichosis. Altrovandus dubbed them the Ambras family, portraits of the family was found as evidence. During the next 300 years, about 50 cases were observed. The scientist Rudolf Virchow described a form of hypertrichosis accompanied by gingival hyperplasmia in 1873

Figure 1: ambras syndrome

Curse upon "julia pastrana "

Julia pastrana (1834-1860) was famously known as bearded lady, she roamed all over United States as a freak and she attained the attention of many artist . She is portrayed as a bearded lady

having extensive hairs distributed equally throughout the her body surface , even on her hands .She was suspected to have congenital hypertrichosis lanuginosa ; however the generalized form of the syndrome coupled with gingival hyperplasmia indicated her condition as congenital terminal hypertrichosis. Still the science failed during those period of time , due to is poor development during those time accompanied with myths, This was not confirmed until after her death (150 years ago), when it became clear that her X-linked syndrome resulted in terminal hairs.

The book of world records

In 2011, Supattra Sasupan born on August 5, 2000 was an 11-year-old girl from Thailand with hypertrichosis was named the world's hairiest girl as Supatra Sasuphan, by the "Guinness book of world records".

Genetics rocked the play

One record in history concerning congenital hypertrichosis lanuginosa is the hairy family of Burma, a four-generational pedigree of the disease. In 1826, John Crawford was leading a mission for the Governor-General of India through Burma. He tells of meeting a hairy man, Shwe-Maong. Shwe-Maong lived in the court of King Ava and acted as an entertainer. Shwe-Maong had four children: three unaffected children, and one child with congenital hypertrichosis, named Maphoon. On a second mission to Ava, Maphoon was described as a thirty-year-old woman with two sons, one of which had hypertrichosis. The affected son was named Maong-Phoset. He had an affected daughter named Mah-Me. whereas all affected members of the family had dental problems; the unaffected members had perfect teeth. The lack of definitive terminology can be confusing and may make the distinction of the related but unique hypertrichosis syndromes difficult. [1-21]

Types (1)

There are several types of hypertrichosis seen they are mentioned below,

Congenital hypertrichosis lanuginosa

It appears as normal laungo (fine hair found on baby at birth), but instead of disappearing during subsequent weeks, the soft fine hair continues to grow in various places on the baby's body

Congenital hypertrichosis terminalis

Abnormal hair growth begins at birth and continues throughout person's life as curse .They is usually long and thick, found all over the body and face.[3]

1. **Nevoid hypertrichosis:** Excessive hair growth appears as a patch on a defined area

2. **Hirsutism:** This is limited to women; it results in dark, thick hair growing in places where women normally don't have hair (such as face, chest and back)
3. **Acquired hypertrichosis:** They tend to develop later in life. Excess hair may grow in small patches.

Etiology

1. Persistent hair growth

Human hair differentiates into lanugo, vellous, and terminal hair.

- **Lanugo hair**: It develops and sheds during fetal life and is immediately followed by vellous hair.
- **Vellus hair:** It is seen over the face and arms of children; it is shorter, softer, lightly pigmented, and may be medullated.
- **Terminal hair**: It is longer, coarser, and medullated; it is found on the scalp, eyebrows, and eyelashes from birth, and it comprises the new hair and the hair pattern formed in puberty and adulthood. Terminal hair arises from vellous hair follicles under the influence of androgens.[5]

2. Reactivation of genes that cause hair growth
3. Malnutrition
4. Mutation
5. Diet or eating disorder like **anorexia nervosa**
6. Drugs (androgenic steroids, the hair growth drug such as minoxidil, cyclosporine)

Pathophysiology

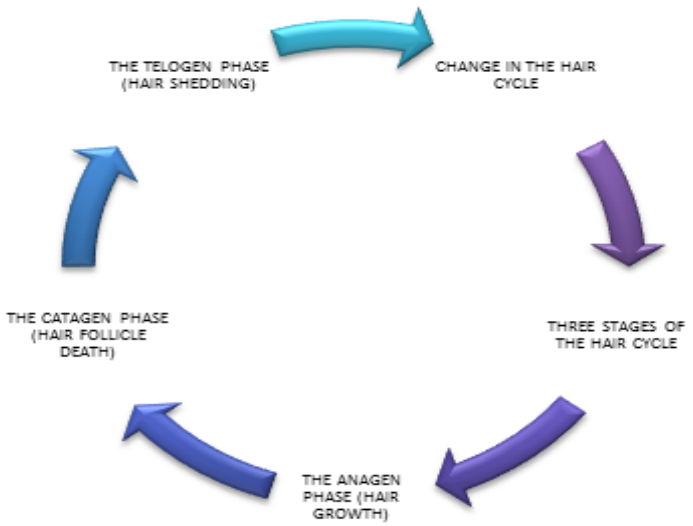

Figure 2: Pathophysiology

- A number of mechanisms can lead to hypertrichosis. One cause involves areas of the skin that are transforming from the small vellus type to the large terminal type.
- This change normally occurs during adolescence, when vellus hair follicles in the underarms and groin grow into terminal hair follicles.[25]
- 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 stages of the hair cycle: the anagen phase (hair growth), the catagen phase (hair follicle death), and the telogen phase (hair shedding).
- If the anagen phase increases beyond what is normal; that region of the body will experience excessive hair growth.
- If the anagen phase increases beyond what is normal. That region of the body will experience excessive hair growth .which results in hypertrichosis.

Prevalence of hypertrichosis in other states

Congenital forms of hypertrichosis are rare. Only 50 cases of congenital hypertrichosis lanuginosa have been recorded since the middle ages and fewer than 100 cases of congenital generalized hypertrichosis have been documented in scientific publications and by the media. Congenital generalized hypertrichosis is isolated to one family in Mexico. Acquired hypertrichosis and Hirustism is much more common, affecting about 7 percent of the female population in the U.S.[26]

Prevalence in INDIA

Nearly 80 percent of the women population was affected by facial hypertrichosis, Due to their hormonal imbalance, and due to their genetic drift. Mensural problem is not taken into account for this issue.

Diagnosis

Hypertrichosis is diagnosed clinically by the occurrence of hair in excess of what is expected for age, sex, and ethnicity in areas that are not **androgen-sensitive**. The excess can be in the form of excessive length or density and may consist of any hair type (lanugo, vellus or terminal hair).

Management

There is no cure for any congenital forms of hypertrichosis. The treatment for acquired hypertrichosis is based on attempting to address the underlying cause. Acquired forms of

hypertrichosis have a variety of sources, and are usually treated by removing the factor causing hypertrichosis, e.g. a medication with undesired side-effects.

Avoiding certain medication such as minoxidil

Depilation methods

- They involve methods like trimming, shaving, and usage of depilators.
- They remove hair to the level of the skin and produce results that last several hours to days

Epilation methods

- They involve methods like plucking , electrology , waxing , sugaring , threading
- It result in the removal of hair from the root, the result last from several days to weeks .

Short Term or Temporary hair removal treatment

- Temporary hair removal may last from several hours to several weeks, depending on the method used.
- These procedures are purely cosmetic.
- They are less expensive.

Removal of hair using by short term methods involves the following

1) Shaving
2) Chemical epilation
3) Waxing
4) Plucking
5) Hair bleaching
6) Sugaring
7) Threading
8) Usage of epilators
9) Applying heavy makeover

Long Term or Permanent hair removal [1]

- Permanent hair removal uses chemicals, energy of various types, or a combination to target the cells that cause hair growth.
- It is an effective method of hair removal on hairs that have color.
- The act on target zone.
- It is very expensive.
- They require additional sessions for the complete removal of hair.

Removal on hair by Long term methods include

1) Electrolysis
2) Laser surgery

Laser Technique

- It is an effective method of hair removal on hairs that have color.
- Laser cannot treat white hair. The laser targets the melanin color in the lower 1/3 of the hair follicle, which is the target zone.

Food and Drug Administration (FDA)

- The U.S. (FDA) allows only electrology to use the term "permanent hair removal" because it has been shown to be able treat all colors of hair.
- The practice of electrology was an absolute victory.
- They improve the self-esteem of the patients.

Science the wonder medicine

- Science paves the way for the improvement and identification of alternate remedies for all aliments, it involves the following identifications of medicines for hypertrichosis;
- Medications to reduce the production of hair were currently under testing.
    i. One medication involves in the suppression of testosterone by increasing the sex hormone-binding globulin.
    ii. Another controls the over production of hair through the regulation of a luteinizing hormone.

Advantages of management

1. They will provide complete cure
2. They ensure positive thoughts among the population
3. Improves self-esteem in person
4. Doesn’t damages the epidermal layer of the body
5. Effective for removing hairs from large areas such as back and legs
6. Complete reduction is observed

Disadvantages of management

1. Highly expensive
2. Painful process
3. May cause burns
4. Time consuming process

5. Adverse reactions are seen in case of medicines
6. May cause irritation and itching of skin
7. Regrowth of hair is seen after weeks to month
8. Development of skin cancer is noticed after the administration of heavy laser rays.
9. Temporary color changes in the area of treated skin are observed for laser therapy.
10. Rarely purple coloring of skin is seen

CONCLUSION:

The disease which was criticized by the historical myth of the early people now came out with the full pledged medical condition known as hypertrichosis. there is no complete eradiation of the disease thou many cosmetical alternatives are available as the result of developing science in the field of medicine and cosmeticology, the mindset of people provide a wide range of improvement in science.

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