

Description of the case of monilethrix in 3 years old child

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Case report

The patient 3-year old girls with complaints on the hair loss, fragility of hair, inability to grow hair the desired length. The first signs of the disease showed up at the age of 1-2 months when, after the loss of vellus hair was observed growth retardation of new hair that caused concern at the parents. The new hair differed dryness and fragility. Grow hair longer than 2cm could not. The child was fourth by count in the family. Family history is not burdened, in both parents and relatives the violation of hair is not have been observed. Data about the disease in distant relatives are unknown. The development of child corresponds to the age, somatically healthy.

Status praesens communis: hair on the head short, with the different length (0,5-2 cm), dry, fragile, uneven painted on the back of the head and back side of the neck. Can be seen small nodules hyperkeratosis.

When microscopy on the rods hair determined by the spindle - shaped thickening and waist.

It was established diagnosis - monilethrix. Recommended: products with high iron content, vitamins A and E, gelatin, multivitamins with age dosage, locally on the occipital area of lotion with 5% urea 2 times per day, gentle care of the hairy part of the head with the exception of slip and trauma, observation by the thrihologist.

Monilethrix (monilethrix; lat. "monile" necklace + Greek "thrix" hair) of genetically deterministic pathology hair, manifested an anomaly hair of the rod, accompanied by its fragility caused by the structural weakness of hair fiber in hair keratins. The hair aplasia like a spindle was first is described in 1879 by Lewis and Smith.

The most common autosomal-dominant type of inheritance, it causes because of the mutations in keratins number 81, 83 and 86. All these keratins are specific for hair shaft. The autosomal-recessive monilethrix caused by the mutation in desmosomal cadgerreen DSG4 is less common [1-4].

This pathology can develop as in women, as well as in men, but more often suffer woman.

Clinical signs

The main complaints of monilethrix are the loss and thinning hair on the head or on other areas of the body of different severity, the presence of small follicular papules of hyperkeratosis are often throughout the surface of the scalp. If you touch the hair its like "grater" on the back of the head and rear surface of the neck nodules more and they have a reddish pink color. Hair short, broken off at the level of 1-2 cm, dull, dry, deformed, non-uniformly dyed, have a spindle- shaped swelling and waist. The defeat of the hair can be isolated or diffuse, more often more expressed in the occipital area. At the microscopic

study shows that on the broad areas of hair shaft has a normal structure, and in the thin places found thinning of cortical layer for because of reduce and dystrophy its cells. The cells deformed with the formation of outgrowths as a comb. The breaking of the hair takes place in places of thinning.

Histology

There are identified horny plug in the mouths of hair follicles, deformation of hair follicles, hyperplasia granular layer, acanthosis and spongiosis. It is noted dystrophy neuro- receptor apparatus.

The disease can exist with the birth, but more often manifests itself on the first year of life, when the fall out vellus hair, and the normal do not grow or appear broken. Extremely rare disease manifests in a later childhood and still less likely - in adults.

The prognosis of this disease is unknown. The disease can exist without changing throughout life, some improvement of hair is celebrated in the summer, in adolescence, as well as during pregnancy, but most often characterized by gradually progressive for when on the site of follicular papules is developing atrophy. In rare cases with age may spontaneous cure.

General condition of patients usually do not suffer.

However, monilethrix can be combined with:

- dystrophy onail(thickening or thinning of the nail plate, it's deformation or complete absence of nail);
- a lag in the mental and physical development;
- two way juvenile cataracts(developed for 2- 5, the life, syndrome Saburo);
- follicular keratos of limbs(syndrome Rața- Turi);
- anomalies of amino acid exchange.

Differential diagnosis

Diagnosis is composed of the analysis of clinical where the indicative is the state of the scalp with follicular hyperkeratosis, as well as research hair under a microscope (structure of the hair like a necklace - characteristic sign of monilethrix). Differentiate need to the trichotillomania, pili torti, fungal diseases, nodosum fragility of the hair and pseudomonilethrix. Pseudomonilethrix the most similar to this disease, but it is develops only in boys 8-14 years old and do not

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shown hyperkeratosis on the scalp.

Treatment

There is no specific and effective treatment.

Drugs and procedures which facilitate course of the disease:

- orally - the vitamins A, B5, F, E, preparations containing phytin(calcium magnesium salt phytic acid), iron and food gelatin;

- the local treatment - softening keratinized particles skin, exfoliating and mitigating ointment(1% salicylic, ointment with 5% urea);

- treatments with irritating effect on the skin (cryomassage, electrophoresis, massage of collar zone);

- if it necessary - sedatives and hormonal means.

Conclusion

Monilethrix has a hereditary nature of the long- and unpredictable course. Is important advocacy with parents in relation to the rules of care of hairy part of the head and smoothing of adverse effect of cosmetic defect on the psyche of the child.

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