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Coexistence of Woolly Hair and Monilethrix: A Cases Study

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Abstract: Both Monilethrix and Woolly hair are considered as a rare autosomal dominant disorder with variable penetrance. Woolly hair exhibits short, kinked hair, which may also comprise the hair over the other places of the body. While Monilethrix is characterized by fragile hair that causing in patchy dystrophic alopecia. Here, we report a case of two sisters with an abnormality of scalp hair characterized by shorter, dry, finer light brown, kinky with a tangled appearance throughout the scalp. Trichoscopic and microscopic findings achieved the diagnosis. A therapeutic application with topical minoxidil was performed.

Keywords: Diffuse Woolly Hair, Monilethrix Like Hair, Topical Monixidil

INTRODUCTION

Moniletrix, as a term, derives from the Latin word monile (necklace, in Latin) and thrix (hairs, in Greek). It is a rare hereditary disorder caused by mutations encoding trichocytes, hHb1, hHb3 and bHb6 type-2 keratins, located on chromosome 12q11q13 (Ferrando et al., 2012). Patients with this disorder exhibited elliptical nodes and intermittent constrictions on the hair shafts, causing concise, fragile hair emerging from keratotic follicular papules, particularly in the occipital area (Haliasos et al., 2013). In mild cases, the clinical appearance may be unseen with limited affected follicles. However, in severe conditions, the secondary sexual hairs and the eyebrows and eyelashes may be compromised (Ferrando et al., 2012).

Woolly hair is also a rare congenital structural irregularity of scalp hair displaying as short, kinked hair associated with keratosis pillars. It can also be exhibited in Naxos or Carvajal syndromes. The hair shaft in woolly hair cases shows an elliptical cross-section, axial rotation and kink construction. Woolly hair can be categorized into; the autosomal dominant woolly hair, also termed hereditary woolly hair, and the less frequent autosomal recessive familial woolly hair (Vasudevan et al., 2013).

We herein report two cases presented with the coexistence of woolly hair and monilethrix like hair. Such a combination of results has not been previously reported in the literature.

Cases Summary

Two sisters of non-consanguineous parents aged 1.5 and 11 years old presented with abnormal hair from 6 months. The two children were born with normal black hair all over the scalp. Subsequently, the hair of both cases was gradually changed from dark to light in colour with a dry, sticky like appearance (Figure 1). Their parents noticed that the new hair did not grow long eyebrows had sparse hair. The two sisters' developmental milestones were not delayed, and there was no physical or mental retardation. There was no family history of hair disorder or any other cutaneous lesions.

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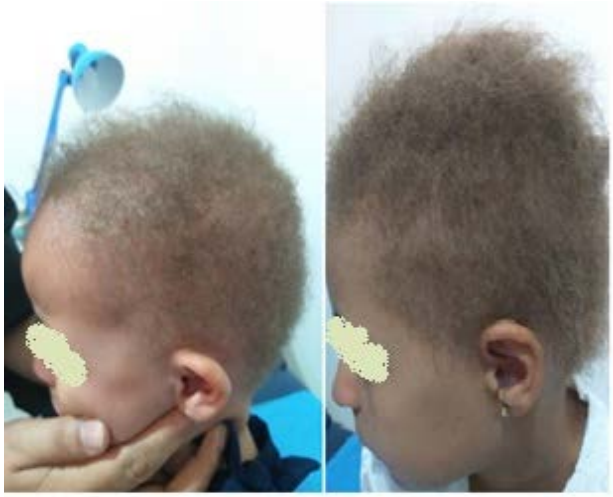
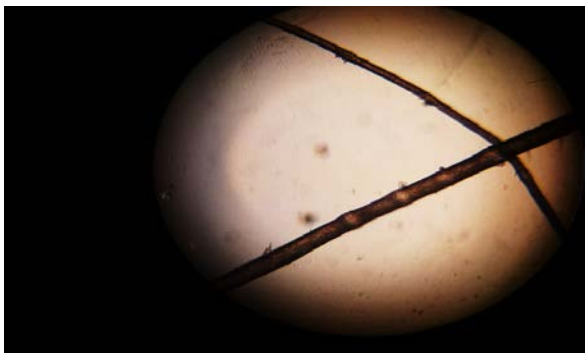


Figure (1). Woolly hair on the scalp with monilithrex like hair

Clinical examination of these cases shows dry sticking hair that could not be combed flat and lighter in colour (blond to light brown). There were no nails and dental, or other skin changes. Laboratory investigations revealed normal haematological and biochemical parameters. Light microscopy and trichoschopy of the uncombable hair showed crawling snake kinging of hair shaft 180 degrees along its axis, and monilethix like hair resembles a string of evenly-spaced beads and non-specific changes such as features of trauma trichorrhexis nodosa (figure 2-4).Scanning electron microscopy was not performed as it was not available in the clinic. Therapeutic strategy with oral zinc syrup 50 mg twice daily and minoxidil spray 2% were started. The two cases and their parents were counselled about the benign and chronic nature of the condition, and were advised for a regular clinical visit.



Figure(2). Microscopic features of monilitherx hair beaded hair shaft and trichorrhexis nodosa

Figure (3). Microscopic features of Woolly hair showing kinking of hair shaft 180-degree longitudinal twisting

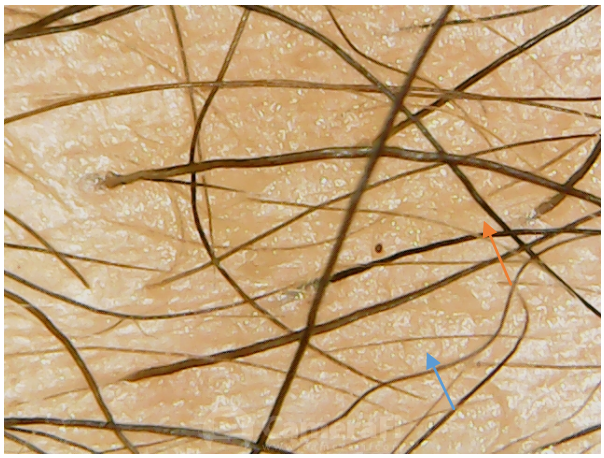


Figure (4). Dermoscopic aspect showing a-crawling snak appearance (Blue arrow monilithrex hairs (red arrow).

DISCUSSION

As stated by Gossage in 1907, Woolly hair is a rare congenital irregularity of the structure of scalp hair.(Gossage, 1908). It is enormously curly, with the typical hair of 0.5 cm in diameter. Woolly hair can appear as a part of systemic disease (known as Woolly hair syndrome) or occur without any systemic outcomes (non-syndromic woolly hair). Woolly hair can be categorized into four types: hereditary, familial, symmetrical circumscribed allotrichia, and woolly hair nevus (Hutchison et al., 1974).

In addition to woolly hair, the patient in this research had Moniletrix disorder, an autosomal-dominant disorder with inconstant pene-

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trance. Several treatment strategies such as griseofulvin, retinoids, topical 2% minoxidil, and oral N-acetyl cysteine have revealed provisional and revocable improvement in such cases.Congenital structural woolly hair has been previously described in many generations of a German family. Autosomal dominant woolly hair has also been stated in an English family with related ichthyosis and deafness (Verbov, 1978). Some other features were presented in a Dutch family with woolly hair involved acral keratoderma, dental caries, and facial irregularities (Van Steensel et al., 2001).

A case of woolly hair with Moniletrix irregularities has not previously been reported in the literature. However, cases of woolly hair with other anomalies such as; spinulosa decalvans, keratosis follicularis, scarring alopecia, teeth and ophthalmological irregularities has been stated (Van Steensel et al., 2001). Additional cases of woolly hair accompanying pachyonychia congenita, nail changes in the form of yellowish-brown hypertrophy and wedge-shaped subungual hyperkeratosis of all 20 nails with birth teeth have been conveyed (Ehsani et al., 2008).

CONCLUSION

To the best of our knowledge, woolly hair with the coexistence of monilithrex like hair findings has not been described previously in the literature.

ACKNOWLEDGEMENT

Declaration of patient consent, all appropriate patient consent forms were obtained and financial support and sponsorship were nil**.**

ETHICS

There are no conflicts of interest.

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تزامن وجود الشعر الصوفي، ومونيلثريكس: دراسة حالة

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المستخلص : يعد كل من شعر Monilethrix ، وWoolly من الاضطرابات السائدة والنادرة في الشعر ذات الاختراق المتغير، يظهر الشعر الصوفي على هيئة شعر قصير مجعد، والذي قد يشتمل أيضًا على الشعر فوق أماكن أخرى من الجسم. بينما يتميز Monilethrix بالشعر الهش الذي يسبب ثعلبة ضمور غير مكتملة. تتناول الدراسة حالة أختين لديهما شذوذ في شعر فروة الرأس يتميز بقصر، وجفاف، ولون بني فاتح، وله مظهر متشابك في جميع أنحاء فروة الرأس. حيث تم التوصل إلى التشخيص من خلال النتائج المجهرية، ومنظار الشعر، وتم إجراء تطبيق علاجي مع المينوكسيديل الموضعي.

الكلمات المفتاحية: شعر صوفي منتشر ، مونيليثريكس مثل الشعر ، مونيكسيديل موضعي.

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