

Woolly hair nevus type 2 : A case report

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ABSTRACT:

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Woolly hair nevus is a rare, nonhereditary disorder of the hair shaft. Approximately 50% of the patients with woolly hair nevus are associated with epidermal nevus (woolly hair nevus type 2). Other cutaneous and extracutaneous lesions have been reported in association with this condition. In this report, we describe a patient with woolly hair nevus type 2. A 52-year-old woman presented with fine, curly, woolly-textured hair on top of verrucous, hyperpigmented plaques. Scanning electron microscopic and histopathological findings are compatible with woolly hair nevus type 2. There is no other systemic manifestation found.

Key words: woolly hair nevus type 2, woolly hair associated with linear verrucous epidermal nevus.

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บทคัดย่อ:

จันทกานต์ นิตินาวรัตน์, รัชต์ธร ปัญญาประทีป, ภาวินี อุทชนิมิตร รายงานผู้ป่วยโรค WOOLLY HAIR NEVUS TYPE 2 วารสารโรคผิวหนัง 2558; 31: 187-192.

สาขาวิชาตจวิทยา ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย

Woolly hair nevus เป็นโรคที่มีความผิดปกติของเส้นผมชนิดหนึ่งที่พบได้ยากและไม่ถ่ายทอดทางพันธุกรรม ประมาณครึ่งหนึ่งของคนไข้จะพบ epidermal nevus ร่วมด้วย (ถูกเรียกว่า woolly hair nevus ชนิดที่2) มีรายงานความผิดปกติอื่นร่วมด้วยได้ทั้งที่ผิวหนังและระบบอื่นๆในคนไข้ที่เป็นโรคนี้ รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยหญิงอายุ 52 ปี มาตรวจรักษาด้วยเรื่องกลุ่มผมเส้นเล็กหยักคล้ายขนสัตว์บนผิวนูนหนาที่ศีรษะ ผลตรวจทางจุลทรรศน์อิเล็กตรอนแบบสแกนนิ่งและจุลพยาธิวิทยาเข้าได้กับ woolly hair nevus ชนิดที่2 การตรวจเพิ่มเติมไม่พบความผิดปกติของระบบอื่นๆ

คำสำคัญ: เนื้องอกของกลุ่มเส้นผมหยักชนิดที่2, เนื้องอกของกลุ่มเส้นผมหยักร่วมกับเนื้องอกของผิวหนังที่เกิดจากการแบ่งตัวผิดปกติของชั้นหนังกำพร้า

Case report

The patient is a 52-year-old woman from Ayutthaya province. Her mother noticed a cluster of hair on her right parietal area to be different from the other areas since her infancy. During her childhood, she developed multiple plaques on that particular area of her scalp, left arm and left leg. She was diagnosed with hypertension and dyslipidemia for 10 years and migraine for 35 years. None of her family members show similar hair abnormality.

Skin examination (figure 1) revealed a well circumscribed area of fine, curly, woolly-textured hair on the right parietal area. The hair elsewhere on the scalp and body normal. There linear, verrucous and hyperpigmented plaques along the blaschko lines on the

affected area of the scalp, left arm, trunk and left leg. Hyperkeratotic plaque on her left sole was identified. Hair pulling and hair tugging test were negative. Physical examination revealed no significant findings in other systems.

The skin biopsy was done showing papillomatosis and acanthosis of epidermis with elongation of the rete ridge. The transverse section showed an elliptical shape of hair shaft (figure 2). Scanning electron microscopy of the hair shaft showed elliptical transverse section and longitudinal groove along the shaft (figure 3). The electrocardiography, echocardiography and cardiac MRI no abnormality. The diagnosis of Woolly hair nevus type 2 (Woolly hair nevus with associated linear verrucous epidermal nevus) was made.



Figure1 (A) Well-circumscribed area of fine, curly, woolly-textured hair on the right parietal area. (B) Linear, verrucous, hyperpigmented plaques along blaschko lines on that area of the scalp, left arm

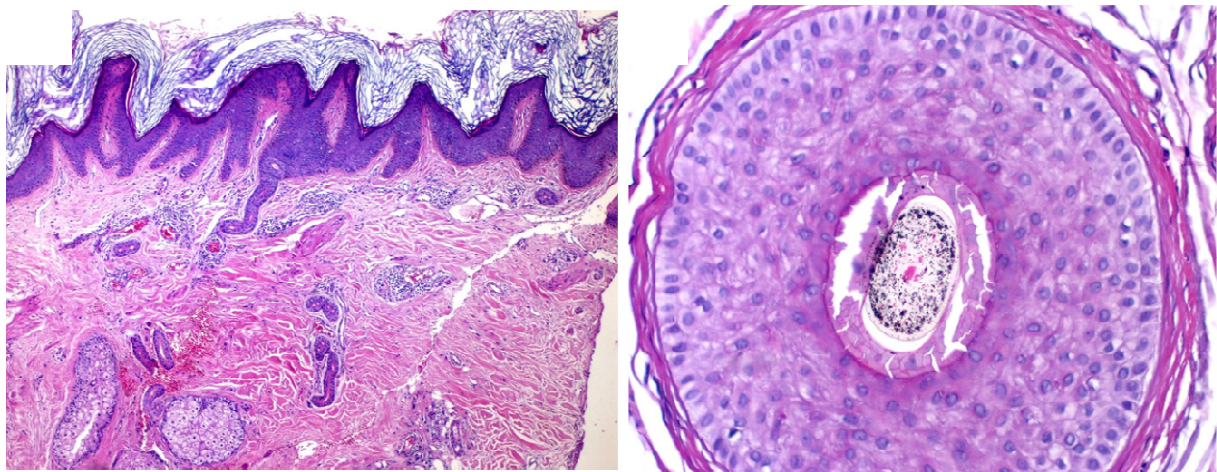


Figure2 (A) Histopathological examination under x100 magnification showed papillomatosis and acanthosis of epidermis with elongation of the rete ridge. (B) An elliptical shape of hair shaft in transverse section taken at x400 magnification.

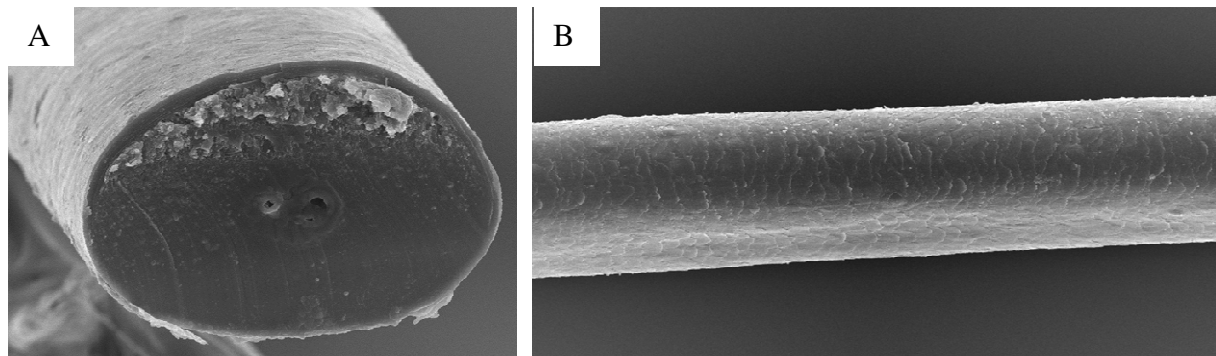


Figure 3 Scanning electron microscopy under x 1500 magnification revealed (A) an elliptical shape and (B) longitudinal groove along the shaft of the hair

Discussion:

Hair shaft disorders are typically classified by the presence or absence of hair shaft fragility. Both congenital and acquired forms are encountered (diagram 1). In this case, the patient presented with hair

shaft disorder without increasing hair fragility. The diagnosis of woolly hair is made from the characteristic hair shaft on physical examination, scanning electron microscopy and histopathology.

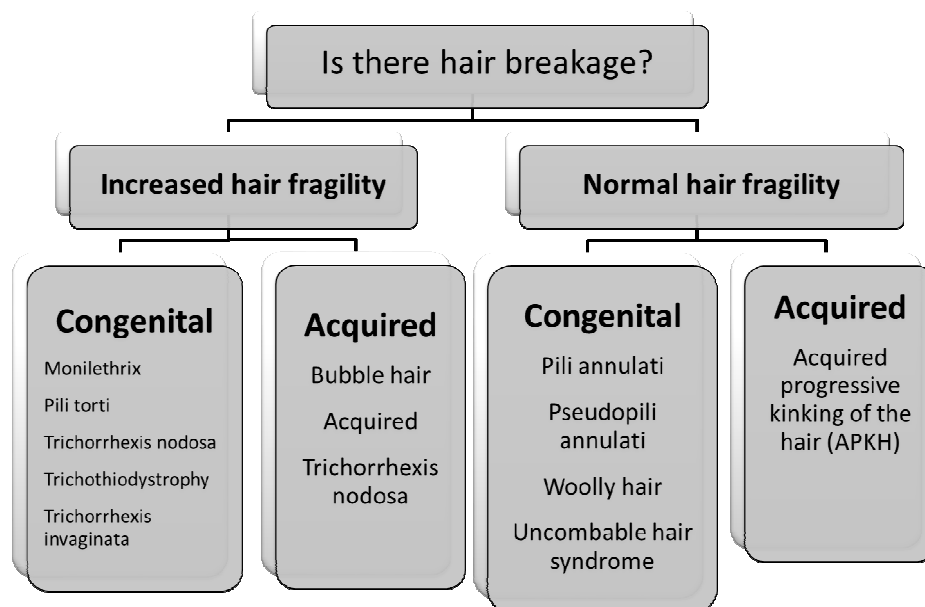


Diagram 1 Approach to hair shaft disorder.

Woolly hair nevus is a rare, nonhereditary disorder of the hair shaft affected one or more circumscribed area of the scalp. Both sexes are equally affected. It is characterized by the presence of extremely curly, woolly-textured, lighter color and thinner hair compared to the rest of the scalp. The onset can be congenital early in the childhood. Approximately half of the patients with woolly hair nevus also have epidermal nevus.¹

Post classified woolly hair nevus into 3 categories.² Type 1 the disease with no association with scalp or glabrous skin disorder. Type 2 disease associated with linear verrucous epidermal nevus that may occur underneath or appeared on other areas of the body. Type 3 the acquired form affected young adult with short, dark, curly hair showing progressive kinking of scalp hair.

There were many cutaneous and extracutaneous anomalies that have been reported in association with woolly hair nevus. These include epidermal nevus; mongolian spots;³ white sponge nevus;⁴ ocular defects such as persistent pupillary membrane,⁵ refraction defect,⁶ retinal defect;⁷ bone growth retardation; tooth anomalies; delayed development of language;⁸ precocious puberty;⁹ and cardiac abnormalities.¹⁰

“Woolly hair nevus syndrome” was used for the first time by Harmozi.⁸ to describe woolly hair nevus type 2 with additional cutaneous or extracutaneous lesions. The summary of case reports of woolly hair nevus syndrome is shown in table1. Therefore, in a patient with woolly hair nevus type 2, it is imperative to search for other systemic involvement.

Table 1 Case reports of woolly hair nevus type 2 with additional finding

Authors	Associated finding	Onset of associated finding	Gender
Legler A ⁴	White sponge nevus	Childhood	Male
Peteiro C ⁶	Refraction defect	Childhood	Male
Harmozi SA ⁸	Bone growth retardation, tooth anomalies, precocious puberty, delayed development of language	6-year-old	Male
Tay YK ⁹	Idiopathic central precocious puberty, multiple acquired melanocytic nevi	20-month-old	Male
Oliveira JR ¹⁰	Conduction defect in the right bundle of his	7-year-old	Male

No effective treatment for woolly hair is currently available. A serial excision of woolly hair nevus is an optional treatment for cosmetic concern. Palmo-plantar keratosis can be treated with moisturizer or mechanical abrasion.

In our case, the studies undertaken to search for systemic involvement revealed no abnormalities. She was advised regarding her condition and received symptomatic treatment.

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