

Case Report

Anterior cervical hypertrichosis associated with speckled lentiginous nevus

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ABSTRACT

Anterior cervical hypertrichosis (ACH), or “hairy throat,” is a rare form of localized hypertrichosis characterized by a tuft of terminal hair on the anterior neck, usually just above the laryngeal prominence. It is usually seen as an isolated finding, but may be associated with systemic disorders such as neurologic abnormalities. We report the case of an 11-year-old girl who presented with ACH associated with speckled lentiginous nevus.

Keywords: Hairy throat, Localized hypertrichosis, Speckled lentiginous nevus

INTRODUCTION

Anterior cervical hypertrichosis (ACH) or “hairy throat” is an uncommon form of regional hypertrichosis that presents with a tuft of terminal hairs on the anterior neck just above the laryngeal prominence. It was first described by Trattner *et al.*^[1] It may be seen as an isolated finding or associated with systemic disorders such as peripheral neuropathy, developmental delay, mental retardation, and ophthalmologic disorders such as optic atrophy and chorioretinal changes.^[2]

Speckled lentiginous nevus (SLN), also known as nevus spilus (NS), is a congenital melanocytic nevus characterized by a lentiginous macule, subsequently developing multiple darkly pigmented macules or papules in a speckled distribution. It may be congenital or acquired.^[3]

CASE REPORT

An 11-year-old girl presented to the outpatient department with complaints of a tuft of hair growth on the anterior aspect of the neck. Her mother noticed it when she was 6 years old. The hair was fine initially, but became longer and thicker with the growth of the child. The child was born out of a non-consanguineous marriage, and there was no history of similar disease in her elder brother or any other family members. There was no history of trauma, inflammation, or any topical applications.

On examination, a tuft of terminal hairs on normal-looking skin was noted just below the laryngeal prominence [Figure 1]. On further examination, an oval-shaped brown macule with an irregular border studded with multiple small hyperpigmented macules was noted on the right arm near the shoulder [Figure 2]. There was no excess facial hair growth, acne, or acanthosis

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Figure 1: Tuft of terminal hair on the anterior neck (arrow).



Figure 2: Nevus spilus on the upper arm (arrow).

nigricans. She had not attained menarche. The child was subjected to a detailed systemic evaluation. Neurological, ocular, musculoskeletal, and Ear, Nose and Throat (ENT) evaluation did not reveal any abnormality. Skin biopsy was not performed due to the mother's refusal. There were no cutaneous signs of occult spinal dysraphism along the posterior midline or in the lumbar region. There were no epidermal or vascular nevi in this child. Based on the history and clinical features, she was diagnosed as a sporadic case of ACH associated with speckled lentiginous nevus.

The patient and the caretaker were reassured about the lesions' benign nature and advised to follow up.

DISCUSSION

Hypertrichosis is the excess growth of terminal hair in any part of the body. It may sometimes result from causes such as hamartomas, topical corticosteroid application, trauma, inflammation, or it may be idiopathic. Hypertrichosis is classified according to the age of onset (congenital or acquired) and the extent of distribution (generalized or localized).^[4] Many genetic syndromes associated with hypertrichosis have been identified. Head-and-neck anomalies are most commonly associated with these syndromes, followed by musculoskeletal and neurological defects.^[5] ACH is an uncommon form of primary regional hypertrichosis with predominant autosomal dominant inheritance. In addition to hypertrichosis, it may present with peripheral sensory motor neuropathy, hallux valgus, optic atrophy, chorioretinal changes, and mental retardation.^[6] Thus, it is strongly advised to take a thorough family history and to perform clinical examinations and investigations in all patients with ACH to exclude possible systemic abnormalities.

Another common location for congenital localized hypertrichosis is the sacral area, the "fawn tail," along the posterior midline. They are important for being a good predictor of underlying defects such as diastematomyelia, meningocele, spina bifida, kyphoscoliosis, or chest deformities. Other similar conditions are hypertrichosis cubiti (Hairy elbow syndrome), which may be associated with low stature or facial asymmetry, and posterior cervical hypertrichosis, which is an autosomal dominant condition associated with kyphoscoliosis.^[4] About 40 cases of ACH have been reported in the literature.^[2] Megna *et al.* reported 77.5% of cases as isolated defects and 22.5% cases associated with other anomalies.^[2] The most common systemic defects are peripheral and sensory neuropathy, mental retardation, and developmental delay. The presence of ACH, along with other neurological or pigmentary defects, may point to defects in the common ectodermal origin of these structures, even though a conclusive association could not be established. Due to the severity of systemic defects and relatively high incidence in the reported cases (22.5%), it is recommended that all cases of ACH be screened for such anomalies.

NS is a relatively common cutaneous lesion that comprises a flat tan macule with more darkly pigmented macules or papules within it. SLNs are occasionally part of complex disorders such as SLN syndrome, phakomatosis pigmentovascularis, or phakomatosis pigmentokeratolica.

Our patient presented with ACH associated with SLN, with no other systemic association. There were no vascular or epidermal nevi. No case of ACH accompanied by NS has

been reported in the literature to date. A case report of ACH along with nevoid and whorled hypermelanosis was reported by Saini *et al.*^[7] The association of ACH and SLN may be coincidental or related to defects in common developmental pathways. The reports of ACH have been too few worldwide to adequately study the individual associations. Although localized hypertrichosis is a cosmetic issue, it is distressing to patients and parents. Hence, along with counseling, laser hair reduction, electrolysis, and intense pulsed light can be advised after 18 years of age, to improve the cosmetic outcome.

CONCLUSION

In this report, a case of anterior cervical hypertrichosis associated with speckled lentiginous nevus has been presented. The association in this patient might be coincidental, but thorough physical examination and systemic evaluation is recommended in all patients with ACH.

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