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Case Series

Clinical Cases of Anterior Cervical Hypertrichosis

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ABSTRACT

Anterior cervical hypertrichosis (ACH) may be only an isolated aesthetic finding, but it may also be associated with other underlying conditions, including neurological and ocular disorders. In order to recognize this uncommon entity, it is imperative to provide detailed physical examination and clinical history to exclude the possible associated abnormalities in patients with ACH. In those cases that ACH represents only a cosmetic problem for the patient, laser hair removal is recommended. We hereby presented 2 cases of ACH in two young girls and discussed about all the differential diagnoses and how to rule out associated anomalies. The most useful workout to rule out associated underlying conditions are described, and cosmetic treatment with laser hair removal devices is also commented.

Keywords
Hairy throat syndrome; Nevoid hypertrichosis; Dysraphism; Anterior cervical hypertrichosis; Congenital hypertrichosis.

INTRODUCTION

Anterior cervical hypertrichosis (ACH), or hairy throat syndrome is a rare form of localized hypertrichosis that refers to the presence of a tuft of terminal hair on the anterior neck, mostly placed on the hyoid region.[1] Although rarely reported, its frequency is probably underestimated. Approximately 40 cases of ACH have been reported in the literature until now.

Although it is usually an isolated finding, it may be associated with systemic disorders such as neurologic abnormalities (i.e., intellectual disability), orthopedic and ophthalmologic disorders among other minor defects.

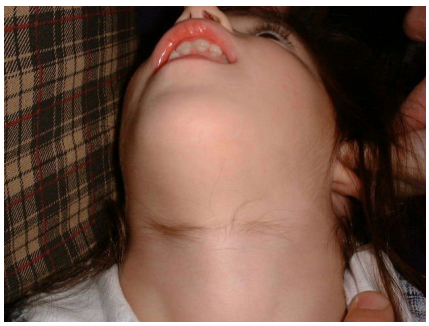
As ACH can be the marker of an underlying defect, it is strongly advised to take a thorough family history and perform clinical examinations and investigations (neurologic and ophthalmologic examination, electromyography, X-ray of the feet) in all patients with ACH to exclude the possible associated abnormalities. We report 2 young girls who presented with isolated ACH.

CLINICAL CASES

Case 1

An 11-year-old previously healthy girl was referred to our clinic for a tuft of blond terminal hair on the hyoid region (Figure 1). Underlying skin had no pigmentary changes. According to parents, the localized hair had started to appear at the age of 4-year and there was no history of swelling in the area, preceding trauma, inflammation, or use of topical drugs. Complete physical examination, including neurological and ophthalmological examination, electromyography and X-ray of the feet, did not reveal any other comorbidities. The patients parents were not consanguineous, and family history was negative for this disorder. Clinical features were consistent with the diagnosis of ACH or hairy throat syndrome.

Figure 1. *Tuft of Terminal Hair on the Hyoid Region of an 11-Year-Old Girl*



Case 2

A 3-year-old healthy girl was seen in our department for the pres-

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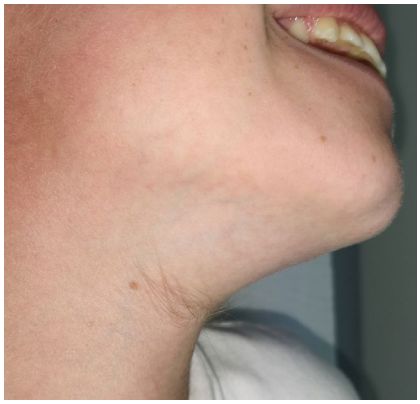
ence of a tuft of hair on the anterior part of the neck (Figure 2). The parents have noticed it from the last year. On physical examination there revealed nothing relevant and neither on personal background. We performed ophthalmological examination and X-ray on feet with no relevant findings. Clinical diagnosis was ACH.

Figure 2. *Hairy Neck Corresponding the ACH in a 3-Year-Old Girl*



DISCUSSION

Anterior cervical hypertrichosis is a rare and little-known form of congenital localized hypertrichosis.[1] It is characterized by the presence of a tuft of terminal hairs in the anterior cervical region.

ACH occurs at birth or during early childhood and it can be acquired or congenital, although the inheritance pattern remains to be determined. Among reported cases, there is a significant female predominance. For most of the affected patients, this anomaly represents only an aesthetic problem; however, in some cases, ACH may be a part of a more complex disorder (22.5% of reported cases). Most commonly associated disorders reported in the literature include neurological abnormalities (with intellectual disability), ophthalmological and orthopedical disorders.[1-6]

In 2015, Megna et al[3] reviewed 40 cases of ACH published to date. The authors concluded that 67.5% cases were familial and 32.5% cases were sporadic. There was a clear predominance on females (75%).

Regarding association with other defects, the authors reported that in 77.5% of cases ACH presents as a solitary defect but in 22.5% is associated with other abnormalities, such as peripheral sensory and motor neuropathy (55% of cases), intellectual disability (22%), developmental delay (22%), ophthalmological disorders (optic atrophy and chorioretinal changes), and hallux valgus.

Differential diagnoses include the following entities:

• Congenital melanocytic nevi; in ACH there is lack of the pigment beneath the hair even though in same cases the pigment might appear later or be rather subtle.
• Dysraphism; usually there is an additional marker like a pit, or a vascular malformation associated with the hair tuff that could mark the defect.
• Nevoid hypertrichosis[7,8]; Nevoid hypertrichosis can be seen in any part of the body and is mostly not associated with any other defect'. Some authors do consider ACH a type of nevoid hypertrichosis.
• Smooth muscle hamartoma; detailed clinical observation and a pseudo-Darier sign may rule out smooth muscle hamartoma.
• Becker nevus; the location of the patch usually helps with (or in) the differential diagnosis of ACH but in early stages the color changes so typical in Becker's nevus might be very subtle.

Regarding treatment options, if there are no associated anomalies, laser hair removal can be extremely useful for cosmetic purposes.[9]

CONCLUSION

Even though ACH might only be a sporadic defect with no other significance than an aesthetic visible defect that can be easily corrected by epilation lasers, in some cases (more than 20%) it might underline an occult abnormality that needs to be ruled out with appropriate diagnostics.

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AUTHOR CONTRIBUTIONS

Dr. Cutrone has provided clinical information from the patients. Dr. Grimalt has written the manuscript. Dr. Valerio has helped on reviewing and editing the manuscript.

STATEMENT OF ETHICS

Written informed consent was obtained from the parents of both patients for publication of this case series and any accompanying images.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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