

CASE REPORTS

Isolated Anterior Cervical Hypertrichosis

B. Monteagudo, M. Cabanillas, C. de las Heras, and J.M. Cacharrón

Servicio de Dermatología, Complejo Hospitalario Arquitecto Marcide-Novoa Santos, Ferrol, A Coruña, Spain

Abstract. Anterior cervical hypertrichosis was described by Trattner and coworkers in 1991. It consists of a «tuft» of hair at the anterior cervical level just above the laryngeal prominence. To date, only 28 cases of anterior cervical hypertrichosis have been reported. Although it is normally an isolated finding, it may be associated with mental retardation, hallux valgus, retinal disorders, other hair disorders, facial dysmorphism, or sensory and motor peripheral neuropathy. We report the case of a 27-year-old woman who presented with this condition as an isolated finding.

Key words: anterior cervical hypertrichosis, localized hypertrichosis, primary hypertrichosis.

HIPERTRICOSIS CERVICAL ANTERIOR ESPORÁDICA

Resumen. La hipertrichosis cervical anterior fue descrita por Trattner et al en 1991. Consiste en la presencia de un «mechón» de pelo a nivel cervical anterior, justo encima de la prominencia laríngea. Hasta la fecha sólo se han descrito en la literatura 28 pacientes con hipertrichosis cervical anterior. Aunque normalmente es un hallazgo aislado, puede estar asociado con retraso mental, *hallux valgus*, cambios retinianos, otras alteraciones pilosas, dismorfismo facial o neuropatía periférica sensitiva y motora. Describimos el caso de una mujer de 27 años con esta entidad como un defecto aislado.

Palabras clave: hipertrichosis cervical anterior, hipertrichosis localizada, hipertrichosis primaria.

Introduction

The term hirsutism refers to the presence of a masculine distribution of hair in women, whereas hypertrichosis is used when there is only an increase in the quantity of hair. Primary hypertrichosis is classified into congenital or acquired, depending on the age at onset, and into localized or generalized, depending on extension. There are 4 conditions included in localized congenital hypertrichosis: hypertrichosis cubiti, anterior cervical hypertrichosis, posterior cervical hypertrichosis, and lumbosacral hypertrichosis.^{1,2}

We present the case of a 27-year-old woman with anterior cervical hypertrichosis, seen in our centre. This is

possibly an underdiagnosed condition, and we have found only 28 cases published to date²⁻¹³; however, it must be taken into consideration in order to exclude associations and avoid laboratory and hormonal studies only necessary in cases of suspected hirsutism.

Case Description

The patient was a 27-year-old Brazilian woman with no past history of interest. She was seen for a cosmetic problem caused by an increase in the hair on the anterior aspect of the neck, present since the first year of life. The patient referred no trauma or inflammation and had not applied corticosteroids or other medication to the area. She did not remember any family history of a similar disorder or of consanguinity.

On physical examination, multiple terminal hairs were observed on normal skin in the anterior cervical region, just above the laryngeal prominence (Figure). Examination of the rest of the skin and skin adnexa was normal, except for mild acanthosis nigricans on the neck and in the axillas.

The diagnosis of anterior cervical hypertrichosis was made based on the clinical findings. Neurological and ophthalmological examinations were normal. The various

Correspondence:
Benigno Monteagudo Sánchez
C/ Alegre 83-85, 3º A
15401 Ferrol, A Coruña, Spain
benims@hotmail.com



Figure 1. Acanthosis nigricans and increased terminal hair in the anterior cervical region, just above the laryngeal prominence.

hair removal techniques were explained to the patient, who chose laser treatment as it is a potentially definitive method.

Discussion

Hypertrichosis is the growth of an increased quantity of excessively thick hair on any part of the skin surface; it can affect both men and women and, in general, there is no underlying hormonal cause. It should not be confused with hirsutism, which is the growth of terminal hair with a masculine distribution occurring in women and sometimes associated with other signs of virilization.^{2,14}

Hypertrichosis is often the result of adverse drug reactions, recurrent trauma, or underlying hamartomas, or

Table. Patients With Anterior Cervical Hypertrichosis and the Associated Abnormalities

Reference	Number of Patients	Familial/Sporadic	Sex	Associated Abnormalities ^a
Vashi et al ²	3	Sporadic	2 M/1 F	No
Trattner et al ³ Garty et al ⁵	3	Familial	1 M/2 F	Peripheral neuropathy (3) Hallux valgus (3) Spina bifida Kyphoscoliosis Optic atrophy Retinal changes
Tsukahara and Kajiji ⁴	7	Familial	3 M/4 F	Turner syndrome
Lee et al ⁶	3	Familial	1 M/2 F	No
Nanda et al ⁷	6	5 familial (2 families)/ 1 sporadic	6 F	No
Braddock et al ⁸	1	Sporadic	F	No
Monteagudo Sánchez et al ⁹	1	Sporadic	F	No
Heitink et al ¹⁰	1	Sporadic	F	No
Thienpont et al ¹¹	1	Sporadic	F	Mental retardation Facial dysmorphism Obesity Hypermetropia Low hairline on back of neck Lumbosacral hypertrichosis
Corona-Rivera et al ¹²	1	Sporadic	M	Mental retardation Abnormal EEG Microcephaly Hallux valgus Inverted nipple Dorsal hypertrichosis Synophrys
Ardinger ¹³	1	Sporadic	M	Peripheral neuropathy Delayed development
Monteagudo et al (present case)	1	Sporadic	F	No
Total	29	18 familial (5 families)/ 11 sporadic	9 M/20 F	22 No 7 Yes

^aThe number of patients affected is specified when there is more than one. Abbreviations: EEG, electroencephalogram; F, female; M, male.

it may form part of a number of syndromes. However, there is a series of primary hypertrichoses classified into congenital or acquired according to the age at onset and into localized or generalized according to the extension. In general, localized congenital hypertrichosis shows autosomal recessive inheritance, is not associated with other abnormalities, and mainly gives rise to cosmetic problems. Four different conditions have been described:

1. Lumbosacral hypertrichosis (faun tail) is the most common. It is present at birth and can coexist with other abnormalities of the skin in this region, such as hyperpigmentation, lipomas, hamartomas, or vascular malformations. It is often a marker of spinal dysraphism, hence the importance of early study to avoid possible neurological sequelae.

2. Hypertrichosis cubiti (hairy elbow syndrome) is observed at birth or during infancy; it is bilateral and, in half of the cases, is associated with a low stature or other malformations such as facial asymmetry.

3. Posterior cervical hypertrichosis is present at birth and has been associated with kyphoscoliosis. Inheritance is autosomal dominant or X-linked recessive.

4. Anterior cervical hypertrichosis.¹⁻³

The aim of treatment in hypertrichosis is cosmetic, and the available methods include bleaching or temporary or permanent methods of hair removal, such as shaving, physical or chemical depilation, electrolysis, and laser or other light sources. Antiandrogen treatments are not used.^{2,6}

Anterior cervical hypertrichosis is a condition described by Trattner and collaborators³ in 1991, and consists of a tuft of hair in the anterior cervical region, just above the laryngeal prominence. Its etiology is still unknown. The absence of underlying abnormalities of the larynx or thyroid indicates that it is not a secondary defect, as in the case of occult spina bifida, for example.¹¹ In general, it has an autosomal dominant inheritance,^{6,8,12,13} although autosomal recessive^{3,6,7} and X-linked dominant inheritance⁴ have also been suggested.

Including our patient, we have only found 29 cases of anterior cervical hypertrichosis described to date, with a predominance in women (20 women to 9 men).²⁻¹³ Eighteen of these patients came from 5 families³⁻⁷ and there were 11 sporadic cases.^{2,7-13} In 22 cases, the abnormality was an isolated finding,^{2,4,6-10} and associated disorders were detected in 7 cases (24%).^{3-5,11-13} Important associated abnormalities reported include neurological abnormalities (abnormal electroencephalogram, peripheral sensory or motor neuropathy, and mental retardation),^{3,5,7,11-13} ophthalmologic disorders (optic atrophy, retinal changes, and hypermetropia),^{3,5,11} hallux valgus,^{3,5,12} kyphoscoliosis,^{3,5} spina bifida,^{3,5} facial

dysmorphism,¹¹ microcephaly,¹² low stature,⁴ synophrys,¹² lumbosacral or dorsal hypertrichosis,^{11,12} and Turner syndrome⁴ (Table). The proportion of patients with abnormalities is possibly overestimated due to the tendency to publish these associations and due to the underdiagnosis of the condition as an isolated defect.⁷ It is unclear if many or all of these associations are fortuitous, but some authors recommend radiological and neurological studies in all patients with anterior cervical hypertrichosis and in their relatives.¹²

Conflicts of interest

The authors declare no conflicts of interest.

References

1. Yuste Chaves M, Zafra Cobo MI, Martínez de Salinas A, Bravo Piris J. Hiperttrichosis cubital. Descripción de dos casos. *Actas Dermosifiliogr.* 2007;98:719-21.
2. Vashi RA, Manzini AJ, Paller AS. Primary generalized and localized hypertrichosis in children. *Arch Dermatol.* 2001;137:877-84.
3. Trattner A, Hodak E, Sagie-Lerman T, David M, Nitzan M, Garty BZ. Familial congenital anterior cervical hypertrichosis associated with peripheral sensory and motor neuropathy—a new syndrome? *J Am Acad Dermatol.* 1991;25:767-70.
4. Tsukahara M, Kajii T. Hairy throat: a dominant trait affecting seven members of a family. *Clin Dysmorphol.* 1992;1:165-7.
5. Garty BZ, Snir M, Kremer I, Yassur Y, Trattner A. Retinal changes in familial peripheral sensory and motor neuropathy associated with anterior cervical hypertrichosis. *J Pediatr Ophthalmol Strabismus.* 1997;34:309-12.
6. Lee HW, Lee MW, Choi JH, Moon KC, Koh JK. Familial anterior cervical hypertrichosis. *J Am Acad Dermatol.* 2005;53:530-2.
7. Nanda A, Al-Arabi I, Ali MT, Alsaleh QA. Anterior cervical hypertrichosis (hairy throat): is it a sign to worry about? *Clin Exp Dermatol.* 2007;32:112-4.
8. Braddock SR, Jones KL, Bird LM, Villegas I, Jones MC. Anterior cervical hypertrichosis: a dominantly inherited isolated defect. *Am J Med Genet.* 1995;55:498-9.
9. Monteagudo Sánchez B, León Muiños E, Cabanillas González M, Martínez Meléndez G. Hiperttrichosis cervical anterior. *An Pediatr (Barc).* 2008;69:591-2.
10. Heitink MV, Quaedvlieg PJ, van Neer FJ, Frank J. Sporadic nonsyndromal anterior cervical hypertrichosis: case report and review of the literature. *Int J Dermatol.* 2007;46 Suppl 3:9-12.
11. Thienpont B, Vermeesch J, Devriendt K. Anterior cervical hypertrichosis and mental retardation. *Clin Dysmorphol.* 2006;15:189-90.
12. Corona-Rivera JR, González-Abarca S, Hernández-Rocha J, García-Cruz D, Corona-Rivera A. Mental retardation in a boy with anterior cervical hypertrichosis. *Am J Med Genet A.* 2005;135:69-71.

13. Ardinger HH. Anterior cervical hypertrichosis versus hairy throat. *Clin Dysmorphol.* 1993;2:186-7.
14. Camacho F. Hirsutismo. Exploración y clasificación general. *Actas Dermosifiliogr.* 2006;97 Suppl 1:1-7.