Case report

GIGANTIC TRICHILEMMAL HORN

M. Bisceglia, A. Bramante, A.L. di Mattia, A. de Cata and T. Paragone

ABSTRACT

A case of a gigantic cutaneous horn of trichilemmal type on the scalp of a young woman is described. Although trichilemmal horns are observed quite frequently, this striking incurved cutaneous horn represents an exceptional example of this entity.

KEY WORDS

trichilemmal horn, giant, case report

INTRODUCTION

Cutaneous horn is a clinical description and not a specific clinical or histopathologic diagnosis. The term cutaneous horn should be applied, when the height of the keratotic mass amounts to at least half of its diameter (2). Cutaneous horns usually are of conical shape and less than one to few centimeters long, being observed in any part of the body, most frequently on the face, followed by scalp, forearms, trunk and genitalia.

We report upon an extraordinary long incurved horn on the scalp of a female patient, which on histopathologic examination revealed on the base of the cutaneous horn multiple, partly opened, trichilemmal cysts.

CASE REPORT

A 55-year-old unmarried, mentally healthy Caucasian woman presented a slowly growing "indurated tumor" on her scalp for almost thirty years, when she was admitted at the Department of General Surgery, "Umberto I" Hospital in S. Marco in Lamis (Italy).

She never complained of pain or any symptoms other than discomfort and private personal feeling of stigma and "shame", letting the horn reach 25 cm in length at the time of the first medical observation (Fig. 1).

For such a long time she hid the horn among her long hair and beneath a dark veil on the head, which she had practically always been wearing during the daytime.



Fig. 1 Gigantic incurved cutaneous horn. The tip is bent downward and inward.

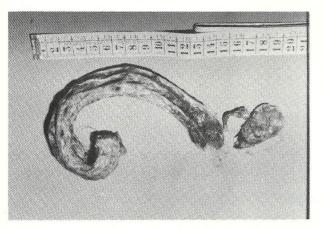


Fig. 2 Gigantic cutaneous horn with base of the surgical specimen.

Shortly after admission, the horn was removed by local exicision with a broad base (Fig. 2).

Histopathologic examination of the base of the biopsy specimen revealed a conglomerate of trichilemmal cysts ruptured or opening into the surface (Fig. 3). The lining of the cysts clearly showed trichilemmal differentiation (Fig. 4).

At the level of its root the horn was composed of orthokeratotic mass surrounded in part by acanthotic epidermis. The patient recovered well and was discharged a week later.

COMMENT

The term "cutaneous horn" refers to a hyperkeratotic skin reaction pattern and not to a specific diagnosis. Verruca vulgaris, solar keratosis, seborrheic keratosis, inverted follicular keratosis, keratoacanthoma and squamous cell

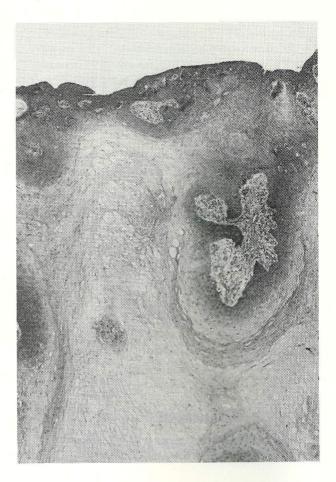


Fig. 3 Scanning magnification shows the base of the cutaneous horn that consists of invaginated keratotic masses surrounded by an epithelial lining exhibiting trichilemmal differentiation. Adjacent to the invaginated epidermis there is a trichilemmal cyst (HE; x10).



Fig. 4 Higher magnification illustrates the trichilemmal differentiation of the epithelial lining of the cyst (HE; x100).

carcinoma are among the most frequent diagnoses thought to be cutaneous horns. Cutaneous horns usually are solitary, though multiple examples have been observed and even in a disseminated form (12).

Less often an epidermal nevus, an open epidermoid cyst and finally an open trichilemmal cyst and a trichilemmoma may represent the primary skin lesion. Trichilemmal cyst and trichilemmoma are practically the only two conditions related to cutaneous horn exhibiting "trichilemmal keratinization" (1, 3, 4).

Trichilemmal keratinization is defined by the production of horny material and keratinized cells without the formation of keratohyaline granules (17). This type of keratinization can be observed in the middle portion of the hair follicle isthmus during anagen and catagen, in the nail matrix and as above mentioned - in trichilemmal cysts (17). Many cases of cutaneous horns are found to be verrucae vulgares on histopathologic examination and some of these cases show trichilemmal differentiation (14). In this context it should be mentioned that it is not yet clear, whether trichilemmoma is a separate entity or a variant of an endophytic wart with trichilemmal differentiation.

Even other different causes in exceptional cases of cutaneous horns have been described, such as Paget's disease (6) and chronic discoid lupus erythematosus (7).

Cutaneous horns are important to dermatologists and pathologists, expecially because they may indicate an

underlying malignancy. The benignancy-to-malignancy ratio has been shown to be 61.1 % vs. 38.9 % in an extensive study (20). The clinical differential diagnosis includes "pseudohorns", i.e. lesions which clinically simulate a cutaneous horn, but histopathologically do not consist of accumulated keratinized material. "Pseudohorns" are composed entirely of benign or malignant neoplastic cells (10, 15, 19, 20, 21), as well as of different organic and inorganic incrustated endogenous or exogenous substances (11, 19). Histopathologically cutaneous horns and pseudohorns may actually represent a problem of recognition and classification, although these lesions are commonly considered as trivial.

The presented cutaneous horn elicits some interest not only because of its extraordinary length, but also because of its remarkable histopathologic findings especially at the base of the horn, namely multiple, partly opened, trichilemmal cysts.

Finally - though the greatest respect should be due to the human dignity - we think this case will be the subject of much curiosity and deserve a place among the oddities of the medical literature, beside the disfigured "elephant" man, affected either by neurofibromatosis type 1 (18) or by the newly recognized Proteus syndrome (5), the "mammouth" tumor, which depicted the largest ever seen abdominal lipoma (8), the "human tails" (9) and few others.

REFERENCES

- 1. Akiyama H, Hagiyama M, Arata J.: A case of trichilemmal horn. Rinsho Dermatol, 1980;22:521-524
- 2. Bart RS, Andrade R, Kopf AW.: Cutaneous horn. Acta Derm Venereol, 1968;48:507-515
- 3. Brownstein MH.: Trichilemmal horn: cutaneous horn showing trichilemmal keratinization. Br J Dermatol, 1979;100:303-309
- 4. Brownstein MH, Shapiro E.: Trichilemmomal horn: cutaneous horn overlying trichilemmoma. Clin Exp Dermatol, 1979;4:59-63
- 5. Cohen MM Jr.: Further diagnostic thoughts about the Elephant Man. Am J Gent, 1988;29:777-782

- 6. Dabski K, Stoll HL Jr.: Paget's disease of the breast presenting as a cutaneous horn. J Surg Oncol, 1985;29:237-239
- 7. Dabski K, Stoll HI Jr.: Cutaneous horn arising in chronic discoid lupus erythematosus (letter). Arch Dermatol, 1985;121:837-838
- 8. Delameter J.: Mammouth tumor. Cleveland Medical Gazette August 1, 1859
- 9. Dao A and Netsky NG.: Human Tails and pseudotails. Hum Pathol, 1984;15:449-453
- 10. Findlay RF, Lapins NA.: Pyogenic granuloma simulating a cutaneous horn. Cutis, 1983;31:610-612

- 11. Jansen T, Kuppers U, Plewig G.: Pomade crust of the scalp simulating cornu cutaneum. Hautarzt, 1991;42:642-644
- 12. Khimkina LN, Radchenko EI.: Case of disseminated primary cutaneous horn with malignant transformation. Vestn Dermatol Venerol Nov (11), 1983;59-62
- 13. Lever WF, Schaumburg-Lever G.: Histopathology of the skin. 7th ed., p.546. Lippincott Company, Philadelphia, 1990
- 14. Nakamura K.: Two cases of trichilemmal-like horn. Arch Dermatol, 1984;120:386-387
- 15. Peterson JI, McMarlin SL: Metastatic renal cell carcinoma presenting as a cutaneous horn. J Dermatol Surg Oncol, 1983;9:815-818
- 16. Pinkus H.: "Sebaceous cysts" are trichilemmal cysts. Arch Dermatol, 1969;99:544-555

- 17. Pinkus H, Iwasaki T, Mishima Y.: Outer root sheath keratinization in anagen and catagen of the mammalian hair follicle. A seventh distinct type of keratinization in the hair follicle: tricholemmal keratinization. J Anat, 1981;133:1-35
- 18. Pomerance B.: The Elephant Man. New York: Grove Press, 1979;1-71
- 19. Rupp SJ, Wagner RF, Porter NC.: Pseudohorn associated with basal cell carcinoma. Cutis. 1991;48:379-380
- 20. Uchiyama N, Shindo Y, Saida T.: Perforating pilomatricoma. J Cutan Pathol, 1986;13:312-318
- 21. YU RC, Pryce DW, Macfarlane AW, Stewart TW.: A histopathological study of 643 cutaneous horns. Br J Dermatol, 1991:124:449-452

AUTHORS' ADDRESSES

Michele Bisceglia M.D., Servizio Department of anatomic pathology, IRCCS Hospital "Casa sollievo della Sofferenza, 71013 S. Giovanni Rotondo (FG), Italy

Antonio Bramante M.D., Department of general surgery, Umberto I., Hospital, San Marco in Lamis (FG), Italy
Antonio Livio di Mattia M.D., same address
Antonio de Cata M.D., same address