A Case of a Proliferating Trichilemmal Cyst With Unusual Clinical Features

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The proliferating trichilemmal cyst is a rare, usually benign tumor of external root sheath origin. About 90% of the cases occur on the scalp, with the residual 10% occurring mainly on the back. More than 80% of the patients are women, most of whom are elderly. The cysts are commonly large, measuring up to 6 cm or more in diameter.

A 33-year-old woman visited our department with a history of a matchhead- sized erythematous tender papule on her left knee that had been present for two years. A histopathological examination revealed a well-circumscribed, multilobulated cystic tumor with trichilemmal keratinization in the dermis. There were individual cell keratinization and focal calcification in some areas.

This case was diagnosed as a proliferating trichilemmal cyst. This case was very unusual not only because the tumor occurred on the knee as a matchhead-sized papule but also because of her young age at presentation. (Ann Dermatol 10:(2) 77~80, 1998).

Key Words : Proliferating trichilemmal cyst, knee

The proliferating trichilemmal cyst, also referred to as a proliferating trichilemmal tumor[1], is a rare usually benign tumor of external root sheath origin[2,3]. This entity has been reported previously under a variety of names such as proliferating epidermoid cyst[4], invasive pilomatrixoma[5], trichochlamydocarcinoma[6], giant hair matrix tumor[7], trichilemmal pilar tumor[8] and giant pilar tumor of the scalp[9]. Pinkus[2] applied the term trichilemmal to the cystic lesion.

It usually develops as a solitary tumor on the scalp of elderly women. Its usual clinical presentation is a subcutaneous cystic nodule that occurs over several years and progresses slowly to a large nodular mass, often after trauma or inflammation[3,10].

We report a case of a 33-year-old woman with a small proliferating trichilemmal cyst on her knee.

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CASE REPORT

A 33-year-old woman had had a small tender papule on her left knee for the previous two years. A physical examination of the skin revealed a matchhead-sized erythematous papule, showing central umbilication on the left knee (Fig. 1). The papule was surgically removed.

Histologically, the tumor was localized in the dermis and consisted of sharply defined multilobulated cysts (Fig. 2). The peripheral cells showed nuclear palisading characteristics on a thick basement membrane. The epithelial cells close to the cystic cavity appeared clear and swollen due to glycogen accumulation and underwent abrupt keratinization without granular cell layer. Breaks in the cyst wall and entrance of the stromal tissue were observed. There were individual cell keratinizations without any nuclear atypia (Fig. 3). In some areas, there was focal calcification (Fig. 3, inlet). Some of the cells contained PAS-positive substances (Fig. 4). It was diagnosed as a proliferating trichilemmal cyst.

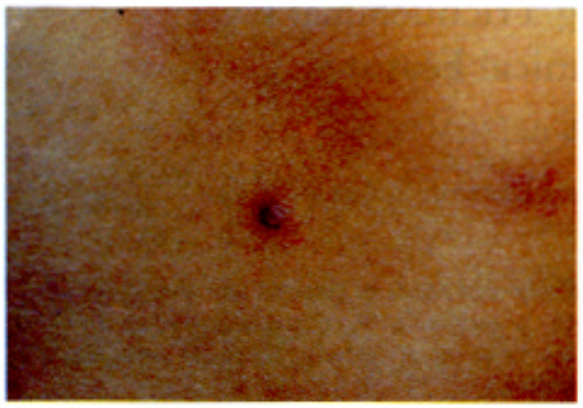

Fig. 1. A matchhead-sized erythematous papule with central umbilication on the knee.

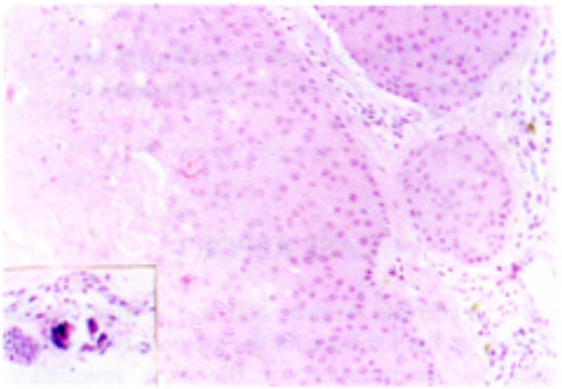

Fig. 3. The cyst walls were composed of squamous epithelium showing abrupt keratinization without a granular cell layer. At the edge of the lobule, cells showed nuclear palisading. There were many individual cell keratinization. Inlet: Focal calcification was observed (×200).

DISCUSSION

The proliferating trichilemmal cyst is a rare benign tumor showing trichilemmal keratinization. Lesions usually present as a solitary, slowly-growing, soft tumors in the deep dermis and often extend into the subcutaneous fat[11].

Histopathologically, the cyst consists of a lobulated intradermal mass of squamous epithelium. Individual lobules are sharply defined and have a clear non-infiltrative border. Peripheral palisading is often present and lobules may be surrounded by a thickened refractile basement membrane. Characteristically, the epithelium in the center of the lobules abruptly changes into eosinophilic amorphous keratin

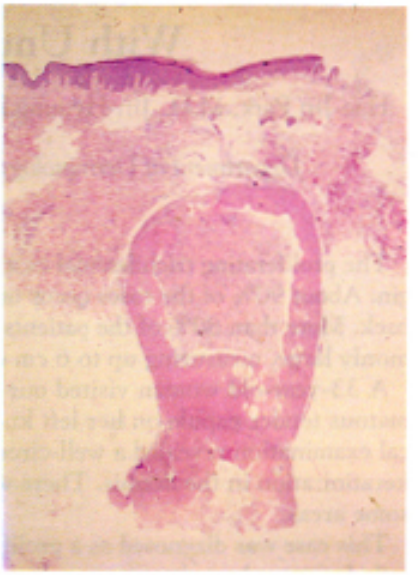

Fig. 2. In the dermis, there is a well circumscribed, multilobulated cyst with a break in the wall (×20).

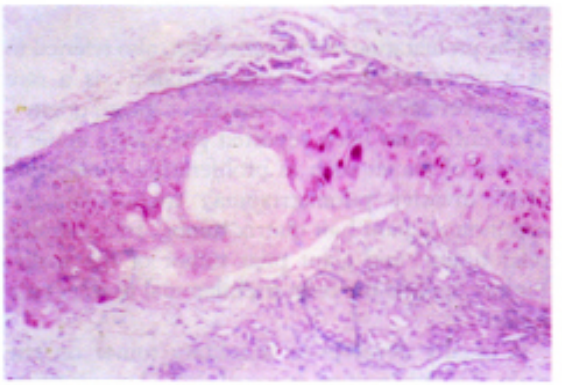

Fig. 4. The clear cells close to the cystic cavity were stained with PAS (×200).

without a granular cell layer. Some proliferating trichilemmal cysts exhibit changes similar to the keratinization of the follicular infundibulum. These changes consist of epidermoid keratinization resulting in horn pearls, some of which are similar to "squamous eddies". In addition, some areas of the tumor may contain a large number of clear cells due to glycogen accumulation[3,11].

In 1968, Holmes[6] showed that the keratin in these cysts or tumors was derived from the outer root sheath of hair. Pinkus[2] firmly established this fact in his paper. The idea that a proliferating

trichilemmal cyst could be a complication of a trichilemmal cyst was proposed by Leppard and Sanderson in 1976[10]. They found that a break in the trichilemmal cyst wall allows the entrance of lymphocytes, histiocytes, giant cells, fibroblasts and small blood vessels. There may be three sequelae to this inflow: 1) the break may be repaired by growth from margins of cysts. Eventually, a new cyst wall may be produced which is nearly indistinguishable from one that has not been damaged; 2) the break may be healed by marsupialization of the cyst; or 3) the lining cells of the cyst may increase in number. This may be the first stage of proliferation that progresses to a pseudoepitheliomatous appearance. This has been mistaken for squamous cell carcinoma. Brownstein and Arluk[3] furthered the notion that a proliferating trichilemmal cyst may occur as a result of trauma or inflammation of a trichilemmal cyst. They indicated that many proliferating trichilemmal cysts had areas indistinguishable from normal trichilemmal cysts and that normal trichilemmal cysts contained areas of hyperplasia identical to proliferating trichilemmal cysts. Furthermore, they found many cysts with a spectrum of changes from minimal hyperplasia to full-blown proliferating trichilemmal tumors. However, Reed[5] and Baptista[12] have suggested that a proliferating trichilemmal cyst constitutes a primary tumor from the onset. In our case, the cyst ruptured and fibrous tissue entered into the lumen. In addition, the lining cells of the cyst showed proliferation and individual cell keratinizations, which is the early stage of proliferation.

Clinically, they are usually large, measuring up to 6 cm or more in diameter[11]. More than 80% of the patients are women, most of whom are elderly. About 90% of the cases occur on the scalp, with the remaining 10% occurring on the back, forehead, wrist and chest[3]. Rare cases occurring on the arm[13] inguinal[14] and vulva[15] skin have also been reported.

This case was unusual not only because the tumor occurred on the knee as a matchhead-sized papule but also because of the younger age at the time of presentation. To the best of our knowledge, there has been no other case that has occurred on the knee.

In rare instances, malignant transformations of a proliferating trichilemmal cyst take place, showing rapid enlargement of the nodule[3]. Jaworski[16] said that the diagnosis of carcinomatous transformations of a proliferating trichilemmal cyst could be made in the presence of abnormal mitoses, marked cellular pleomorphism, infiltrating margins and aneuploidy.

The differentiation of a proliferating trichilemmal cyst from squamous cell carcinoma is one of the most important problems in pathological diagnosis. Features favoring the diagnosis of proliferating trichilemmal cyst over squamous cell carcinoma include areas of trichilemmal keratinization, foci indistinguishable from trichilemmal cysts, calcification, no previous epidermal lesions and sharp circumscription[3].

The treatment of this tumor is surgical excision. It is also important to recognize that it should be excised with a margin of normal tissue because of malignant transformation. Routine follow-up is recommended, especially for the tumors that have a malignant appearance[17].

REFERENCES

1. Hanau D, Grosshans E: Trichilemmal tumor undergoing specific keratinization: "keratinizing trichilemmoma". J Cutan Pathol 6:463-475,1979
2. Pinkus H: "Sebaceous cysts" are trichilemmal cysts. Arch Dermatol 99:544-555,1969
3. Brownstein MH, Arluk DJ. Proliferating trichilemmal cyst: A simulant of squamous cell carcinoma. Cancer 48:1207-1214, 1981
4. Wilson Jones E: Proliferating epidermoid cysts. Arch Dermatol 94:11-19,1966
5. Reed RJ, Lamar LM: Invasive hair matrix tumors of the scalp. Arch Dermatol 94:310-316, 1966
6. Holmes EJ: Tumors of lower hair sheath. Common histogenesis of certain so-called "sebaceous cysts", acanthomas, and "sebaceous carcinomas". Cancer 21:234-248, 1968
7. Dabska M: Giant hair matrix tumor. Cancer 28:701-706, 1971
8. Janitz J, Wiedersberg H: Trichilemmal pilar tumors. Cancer 45:1594-1597, 1980
9. Casas JG, Woscoff A: Giant pilar tumor of the scalp. Arch Dermatol 116:1395, 1980
10. Leppard BJ, Sanderson KV: The natural history of trichilemmal cysts. Br J Dermatol 94:379-390, 1976
11. McKee PH: Pathology of the Skin. 2nd ed. London:Mosby-Wolfe, 15.7-15.9,1996
12. Poiares Baptista A, Garcia E Silva L, Born MC: Proliferating trichilemmal cyst. J Cut Pathol 10:178-187, 1983

13. Yamaguchi J, Irimajiri T, Ohara K: Proliferating trichilemmal cyst arising in the arm of a young woman. Dermatology 189:90-92, 1994
14. Ha SK, Won YH, Chun IK: A case of proliferating trichilemmal tumor unusually occurring on the inguinal area. Kor J Dermatol 31:434-438, 1993
15. Avinoach I, Zirkin HJ, Glezerman M: Proliferating trichilemmal tumor of the vulva: Case report and review of the literature. Int J Gynecol Pathol 8:163-168, 1989
16. Jaworski R: Malignant trichilemmal cyst(letter). Am J Dermatopath 10:276-277, 1988
17. Laing V, Knipe RC, Flowers FP et al: Proliferating trichilemmal tumor: Report of a case and review of the literature. J Dermatol Sur Oncol 17:295-298, 1991