Proliferating Trichilemmal Tumors On Breast And Scalp: Report Of A Case

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Abstract

Proliferating Trichilemmal Tumor (PTT) is a rare but morphologically distinct tumor that usually arises on the scalp of elderly women. It is composed of multiple cysts consisting of squamous epithelium with trichilemmal keratinization without granuler layer interposition. We describe a 47 year-old woman with proliferating trichilemmal tumor one on the breast and four on the scalp.

INTRODUCTION

Proliferating trichilemmal tumor (PTT), is a large, solitary, multilobulated lesion of the hair follicle occurring on the scalp, usually in middle-aged or older women; it is composed of large follicular sheath cells with cytoplasms and is often confused with squamous cell carcinoma. The tumor preferentially arises in areas of dense hair follicle concentrations, and about 90% of cases occur on the scalp, with the residual 10% occurring mainly on the back (1,2). Other, less common locations include the vulva(1,3,4), nose, mons pubis, buttock, wrist, chest, breast(5), upper eye lid(6), dorsum of hand(7), arm(8), organoid nevus(9), shin(10) and elbow(1). Women are affected in more than 80% of cases, and the average age of patients is 65 years (1,11,12). The most characteristic histologic feature of PTT is trichilemmal keratinization. Peripheral cells enlarge and become palestaining and glycogen-rich, with abrupt transition to a dense keratin without the presence of a granular layer $\binom{13,14}{14}$.

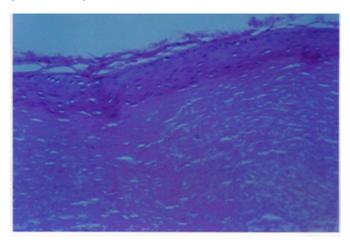
CASE REPORT

A 47 year old woman was admitted to our clinic complaining with mass on breast and scalp. In this report, we describe this patient, who has taken treatment at our clinic for one left breast and four scalp nodules, measuring from 2 to 3 cm in diameters. The mass over the breast was present till 1978. It exhibited slow growth for 25 years and increased slowly in size. The cysts on the scalp were present till 1983. They also exhibited slow growth for 20 years, and increased slowly in size. There was no any sign of pain, erythema and secretions.

The masses on the scalp were present for 10 years. On physical examination, at palpation the mass was diagnosed at upper inner quadrant of left breast, approximately 2,5 cm in diameter, 8 cm far away from areola, mobile and rough. There was not any axillary lymph node. Right breast was normal. There were three rough, non-tender, mobile subcutaneous masses on midline of scalp, ranging from 0,5 cm to 1.5 cm in diameters, and one mass about 0.5 cm in diameter at occipital area, were diagnosed. There was no cervical lymphadenopaty. Laboratory signs were in normal levels. In the cytologic examination of transient needle biopsy (TNB) of left breast; there was inflammatory cells and squamous epithel in area consisting of necrotic material. The subcutaneous nodules were mobile and without an overlying skin ulceration. They thought as trichilemmal tumor. Local excisional biopsy of the masses were performed to the tumors on left breast and scalp, and the pathological diagnosis (Figure-1) returned as Proliferating trichilemmal tumor (Biopsy rapor number:B-3019-03,6853).

Figure 1

Figure 1:Tumor wall and trichilemmal keratinization is seen (H&E:X200).



DISCUSSION

A proliferating trichilemmal tumor (PTT), also referred as proliferating trichilemmal cyst or pilar tumor, is a benign tumor originating from the outer root sheath of a hair follicle (1). It is usually a solitary lesion and most commonlyoccurs in elderly women (13). Although considered biologically benign, PTT may be locally aggressive. In rare instances, malignant transformation has been reported, evidenced by regional or distant metastases (215).

The usual clinical presentation of PTT is that of a long-standing, subcutaneous, cystic nodule that slowly progresses to a large, nodular mass as in our case, and often it follows a history of trauma or inflammation (11,12,13); however, it can also occur de novo, without a preexisting cyst (14). In our case, there were no any history of trauma and inflammation. The presentation is nearly always that of a single lesion but rarely, multiple lesions are seen. Our case was multiple and has seen both on scalp and breast. An associated trichilemmal cyst with lobules of PTT is also encountered occasionally (1). Most investigators now consider PTT as a tumor originating from the outer root sheath of a hair follicle (16,17), and most of the reported cases of squamous or basal cell carcinomas arising in sebaceous cysts are now believed to be PTTs (13,18).

Grossly, PTT appears as a lobulated, well-circumscribed mass, and the external skin may be atrophied or ulcerated. The cut surface generally has a honeycomb appearance with spaces and small cysts filled with keratinous material (13). Histologically, PTT is made up of massively proliferating lobules of squamous epithelium showing multiple central areas of trichilemmal keratinization and formation of

homogeneous keratin cysts (2,12).

Although PTT is generally considered biologically benign, even though histologically indistinguishable from squamous cell carcinoma in some cases, malignant PTT has been reported. Saida et al (19) suggested three stages in the oncologic development of a malignant PTT: the adenomatous stage of the trichilemmal cyst, the epitheliomatous stage of the PTT, and the carcinomatous stage of the malignant PTT. A rare occurrence of PTT with spindle cell carcinoma has also been reported (15). Unfortunately, distinctive histologic or immunohistochemical markers of malignancy do not exist (20). Some recent reports have shown DNA aneuploidy and, in some cases, an increased proliferation index, suggesting that PTT may be a premalignant tumor $\binom{1}{21}$. Rapid enlargement of long-standing nodular scalp lesions and histologic evidence of significant abnormal mitosis, marked cellular pleomorphism, infiltrating margins, and aneuploidy may indicate malignant transformation (1,12). However, the only unequivocal criterion of malignant PTT is metastasis, either regional or distant, and metastases to cervical and mediastinal lymph nodes, lung, pleura, liver, and bones have been reported (2,15).

In our case, there was no any sign of metastasis as a result of all clinical and radiologic scans. There was a study evaluating the PTT, whether it manifests either a cystic or solid mass on imaging studies (22). USG of left breast and scalp revealed a sharply defined mass with spherical and solid echoic lesions. Wide excision of the tumor with a 1 cm conservative margin of normal tissue is the treatment of choice for PTT. Adjuvant chemotherapy is not superior to adequate surgery alone (1). In malignant PTT, the patient should be observed closely to detect any evidence of metastasis.

The importance of our case was the occurrence of proliferative trichilemmal tumors both on scalp and breast. We conclude this case with a review of literature, which had PTT localized on both scalp and breast that excized and diagnosed histologically.

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