GIANT PILAR TUMOR OF THE SCALP

Seyhan ÇENETOĞLU, Serol İNCEOĞLU, Cemalettin ÇELEBİ, Namık K. BARAN, Ömür ATAOĞLU, Tülin OYGÜR,
* From Department of Plastic and Reconstructive Surgery Gazi University School of Medicine, Ankara, TURKEY.
\+ From Department of Pathology,
Gazi University School of Medicine, Ankara, TURKEY.

SUMMARY

GIANT PILAR TUMOR OF THE SCALP

We present a case of a giant pilar tumor of the scalp which has the largest tumor volume ever reported in English literature. The growth pattern and possible role of heredity in such lesions are discussed. The importance of adequate surgical excision and diligent histopathological examination is emphasized.

***Key Word** : Pilar tumor.*

ÖZET

SAÇLI DERİNİN DEV PİLAR TÜMÖRÜ

İngilizce literatürde bugüne kadar bildirilen pilar tümörlerin hepsinden daha büyük hacimli dev bir scalp pilar tümörü sunulmaktadır. Tümörün büyüme özelliği ve kalıtım faktörünün böyle lezyonlardaki muhtemel rolü üzerinde durulmuştur. Tedavide yeterli cerrahi eksizyonun ve titiz histopatolojik incelemenin önemi vurgulanmıştır.

***Anahtar Kelime** : Pilar tümör.*

Pilar tumors are benign neoplasms derived from the outer root sheath of the hair follicle (1). They are believed to have an association with pilar cysts which are sometimes observed to precede or accompany these tumors (2). Although they are known to lack the potential for malignant transformation, evidence of regional metastasis has been shown in several pilar tumor cases in the literature (3, 4, 5). Both because of the locally aggressive growth characteristics and the rare occurrence of regional metastasis, particularly in neglected cases, importance of adequate surgical excision of these tumors has been emphasized previously (6).

In this report, we present the case of an elderly woman with a large, infected and ulcerated scalp mass shown by pathological examination to be a pilar tumor, Personal and family history of the patient were interesting in revealing the role of both budding and multiple cyst formation in tumor growth and pointing out the possibility of inheritance in etiology. This giant pilar tumor (with the largest volume ever reported in English literature) was totally excised and the scalp defect so created was successfully covered with skin grafts. During the three-year follow- up period, no evidence of either local recurrence or regional metastasis was detected.

CASE REPORT

A 78-year old woman who has been suffering from a large scalp mass for 4 years was referred to our deparment with a diagnosis of squamous carcinoma of the scalp. She had first noted a small scalp nodule twenty years ago and later 4 to 5 similar nodules which gradually increased in size and coalesced to from a single mass. Surgical excision had been attempted in the past but failed to cure the lesion. She noted that the surface of the tumor became ulcerated and purulent; foul-smelling discharge and occasional bleeding occured in the last few years. Three of her children, one son and two daughters, both middle-aged, had similar lesions on their scalps. Neither personal nor family history was

otherwise noteworthy. The scalp mass, located in the parietal and occipital regions, had a surface area of 25x20 cm. Its height from the scalp surface varied between 6 to 10 cm. There were foci of ulceration and purulent discharge over its surface (Figure 1).

The cultures from the discharge revealed growth of Staphylococcus Aereus and Pseudomonas Aeroginosa. Her physical examination was normal except for pale conjunctivas. Total blood count and biochemistry showed no abnormality other than mild anemia and hypoalbuminemia. Cranial computerized tomography showed almost uniform invasion of the periosteum and cortical thickening under tumor base but no sign of intracranial spreading (Figures 2 and 3).

Tumor was excised circumferentially with a 1 cm. margin of normal scalp tissue under general anesthesia. Both the invaded and intact appering portions of the periosteum underlying the tumor were included with the excised mass (Figure 4). Burr-holes were formed using a power-drill through the outer table of the skull and capillary bleeding from below was observed.

Three months postoperatively, the whole defect was filled with healthy granulation tissue. Split-thickness skin grafts taken from anterior thigh applied over this tissue provided satisfactory coverage of the defect.

Periodical examinations failed to show any evidence of local recurrence or metastasis in the past three years (Figure 5). The son of the patient was available for excision and pathological examination of his scalp lesion upon his request.

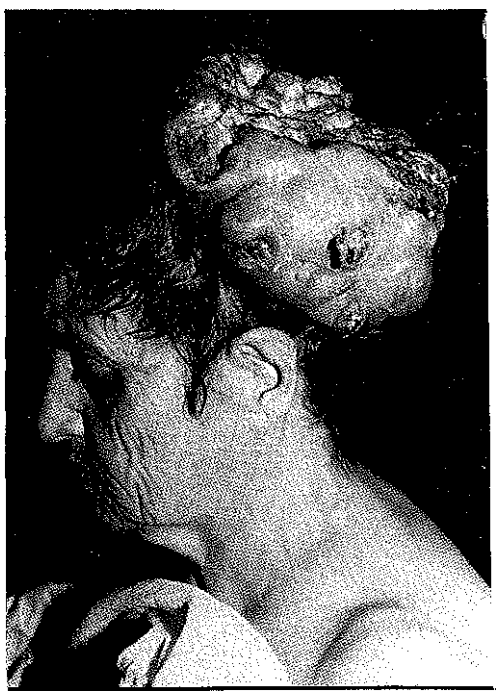

Figure 1: *Lateral wiew of the pilar tumor located in the parietal and occipital regions.*

PATHOLOGICAL FINDINGS

Multiple hematoxyline and eosine sections prepared from the tumoral mass showed lobules of squamous epithelial cells and eosinophilic amorphous keratin in the center of these lobules (Figure 6). Some of the lobules appeared as large cysts filled almost entirely with keratin. The change of the epithelium to keratinous material was abrupt and there was focal calcification in the keratinous material. Epithelial cells showed a slight to moderate degree of atypia and pleomorphism but the mitotic figures were rare; being 1 per 10 high power field in the most active-appearing areas. There was no sign of atypical mitotic figures. Periosteum and bone spicules included with the specimen did not show any sign of tumoral invasion. The lesion excised from the scalp of the patient's son showed typical feature of a pilar cyst.

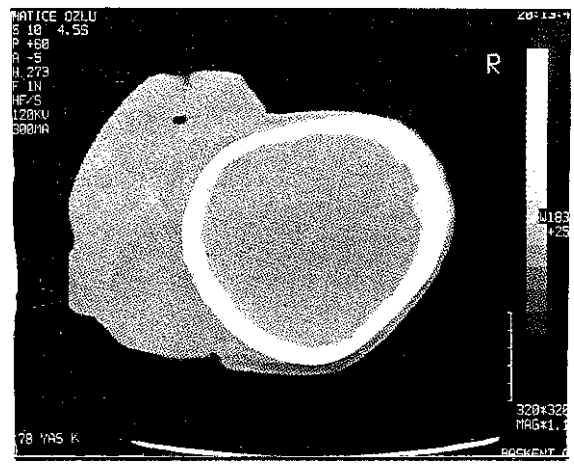

Figure 2: *CT of the cranium showing the huge tumor with no intracranial spreading.*

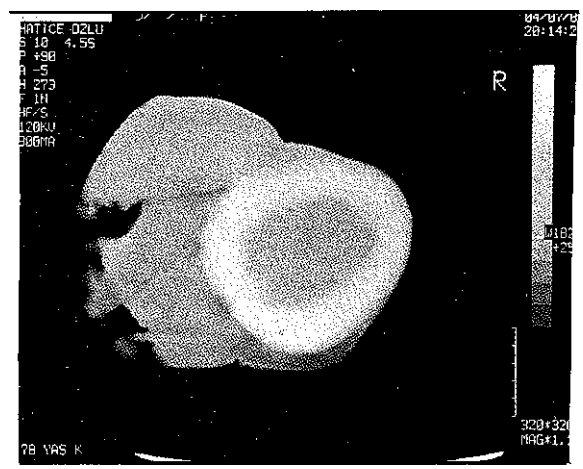

Figure 3: *Another CT section from the same patient showing the ulcerations on tumor surface, periosteal irregularities, and cortical thickening in the skull.*

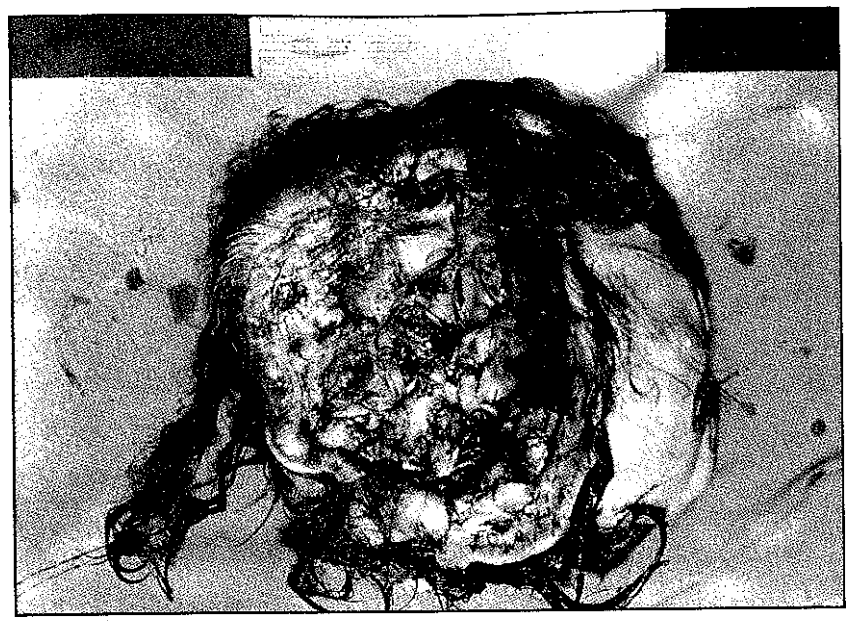

Figure 4: *Photograph of the excised tumor. Note the extensive inflammation and ulceration on its surface.*

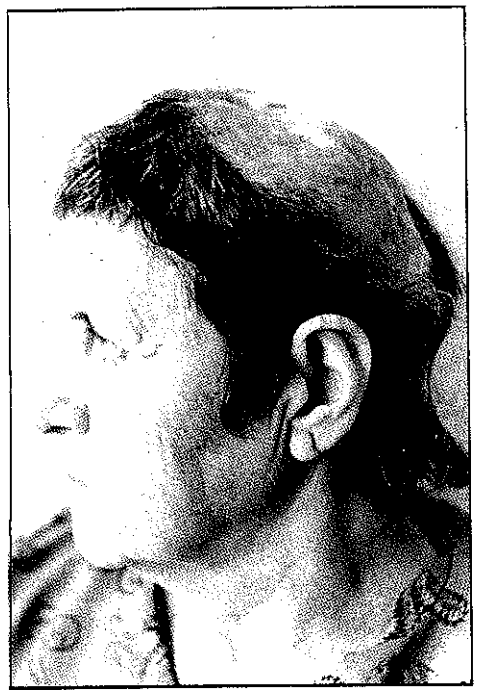

Figure 5: *Two years postoperatively, no evidence of tumor recurrence or skin raft breakdown was observed.*

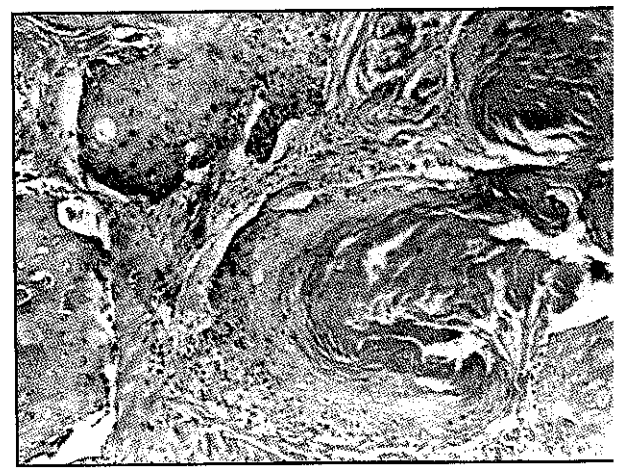

Figure 6: *Lobules of squamous epithelium showing central keratinous material. Note the abrupt change of epithelium to keratin (Hematoxyline and Eosine X 80).*

DISCUSSION

Pilar tumor, which is also called giant hair matrix tumor (7), pilomatrixoma, proliferating trichilemmal cyst or trichilemmal pilar tumor (8), is believed to take origin from the outer root sheath of the hair follicle (1).

The lesion is typically seen in elderly women and is mostly located in the scalp, usually presenting as a solitary cyst or nodule. The size of the cyst increases with time and either budding of the original cyst or occurence of other cysts leads to a mass comprised of miltiple cysts (9). This course has a variable speed and its duration is from a few to over twenty years (8). In this course of events, spontaneous healing or marsupialization and later disappearence of the cyst may be observed. Less frequently, the cyst undergoes proliferation and a pseudoepiteliomathous mass is produced (9). This form of the lesion is likely to be confused with squamous cell carcinoma and, indeed, pilar tumors showing malignant bahaviour have been reported by several outhors (3, 4, 5). This fact points out the significance of a diligent pathological examination for such lesions. Hosokawa et al. (10) and Pinkus (11) have amphasized the importance of histochemical methods used to characterize trichilemmal keratin in lesions such as epidermoid cysts and pilar cysts versus hair matrix. Cotton et al. Showed the difference between the properties of pilar and epidermal cysts using two anti-keratin antibodies derived from two different keratin fractions (12). Various mechanisms of inheritance have been reported for sebaceous cysts and a Mendelian dominant treait has been suggested for a pilar tumor of the scalp that developed in a sebaceous cyst (13). There is no clear documentation of inheritance of de novo pilar tumors. It is interesting to note that the son of the patient in our case had a nodular lesion on his scalp which was excised and confirmed by microscopy to be a pilar cyst.

The pilar tumor presented in this case has the largest volume ever reported in English literature. It is concluded from the patient's history that both budding and separete cystic growth played a role in tumor growth, as was suggested by others (2, 14). Malignant behaviour of the tumor could not be detected by microscopy. Keeping in mind that the lesion recurred twice despite two resections made in the past ten years and reached a huge mass yet retained its benign character, we are convinced that proliferation does not necessarily lead to malign transformation in pilar tumors. The long-lasting inflammation with purulent discharge from the lesion, as observed in this patient, may be a stimulus for this proliferation (1).

Surgical excision with an adequate margin both circumferentially and in deeper layers is the unique treatment for pilar tumors. We believe that the two previous excisions made previously in this case would have been curative had this rule been adhered to. Even in cases where the lesion has invaded the periosteum, the defect created by such extensive resection can be satisfactorily covered with skin grafts.

Seyhan ÇENETOĞLU
Berdan sokak, 4-3,
Kurtuluş, 06600,
Ankara, Turkey.

REFERENCES

1. Baptista. A.P., Silva, L.G.E., and Bron, MC. Proliferating trichilemmal cyst. J. Cutaneous Pathology 10:178, 1983.
2. Hendricks, D.L., Liang, M.D., Borochovitz, D., and Miller, T.A. case of multiple pilar tumors and pilar cysts involving the scalp and back. Plast. Reconstr. Surg. 87:763, 1991.
3. Saida, T., Oohara, K., Hori, Y. and Tsuchiya, S. Development of a malignant proliferating trichilemmal cyst in a patient with multiple trichilemmal cysts. Dermatologica 166: 203, 1983.
4. Batman, P.A. and Evans, H.J.R. Metastasising pilar tumour of scalp. J. Clin. Pathol. 39:757, 1986.
5. Mehregan, A.H. and Lee K.C. Malignant proliferating trichilemmal tumors-report of three cases. J. Dermatol. Surg. Oncol. 13:1339, 1987.
6. Morgan, R.F., Dellon, A.L., and Hoopes, J.E. Pilar tumors. Plast. Reconstr. Surg. 63:520, 1979.
7. Dabska, M. Giant hair matrix tumor. Cancer 28:701, 1971.
8. Brownstein, M.H. and Arluk, D.J. Proliferating trichilemmal cyst: A stimulant of squamous cell carcinoma. Cancer 48:1207, 1981.
9. Leppard, B.J., and Sanderson, K.V. The natural history of trichilemmal cysts. Br. J. Dermatol. 94:379, 1976.
10. Hosokawa, M., Rkugo, S., Aiba, S., Igarashi, M., and Tagami, H. Biochemical characteristics of Keratins from proliferating trichilemmal cyst. Arch. Dermatol. Res. 276:250, 1984.
11. Pinkus, H. "Sebaceous Cysts" are trichilemmal cysts. Arch. Derm. 99:544, 1969.
12. Cotton, D.W.K., Kirkham, N., and Young, B.J.J. Immunoperoxidase anti-keratin staining of epidermal and pilar cysts. Br. J. Dermatol. 11:63, 1984.
13. Strane, M.F., Bennett, M.F. Bennett, M.H., and Melmed, E.P. Pilar tumour of the scalp developing in hereditary sebaceous cysts. Br. J. Plast. Surg. 24:82, 1971.
14. Reed, R. J. and Lamar, L.M. Invasive hair matrix tumors of the scalp. Arch. Derm. 94:310, 1966.