

Proliferating Pilomatricoma Treated Using a Bilateral Advancement Flap

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To the Editor:

Pilomatricoma is a benign neoplasm that is generally straightforward to diagnose clinically. It initially takes the form of a small tumor of nonspecific appearance but displays a characteristic firm consistency to palpation as it develops. We present the case of a giant pilomatricoma on the scalp, noting the abnormal morphology and rapid growth. We also describe a surgical solution in the form of a bilateral advancement flap or H-plasty. The clinical and pathological characteristics suggest that our case could be proliferating pilomatricoma.

Pilomatricoma or calcifying epithelioma of Malherbe was first reported in 1880, when Malherbe and Chenantais described a tumor they initially denominated “calcified epithelioma of the sebaceous gland.” Just a year later, Malherbe published 18 cases stressing the fact that this tumor tended to affect young people. More than 50 years later, in 1933, Chin Luang-Yu identified calcification as another common characteristic of pilomatricoma, and this tumor became known as “calcifying epithelioma of the skin.” In 1942, Turhan and Kranier identified the site of

tumor origin as the hair follicle and shortly afterwards, in 1949, Lever and Griesemer pinpointed it to the primary epithelial germ cells. The current name of pilomatricoma was proposed in 1961 by Forbis and Helwig after they presented a series of 240 examples of the tumor. Subsequent authors contributed further information on the neoplasm: an association with myotonic dystrophy was described by Cantwell and Reed in 1965, and a link with diabetes by Harper in 1971.

We present the case of a 41-year-old man with no relevant medical history, who attended for the appearance of a tumor in the frontoparietal region. Rapid tumor or growth had prompted the patient to make a hasty appointment as the tumor reached the size seen in the photograph (Figure 1) within 4 to 6 weeks. The size of the lesion caused some discomfort and a continual sensation of tension in the area around the tumor that occasionally led to localized pain or a more widespread headache (possibly of a tension type).

Examination revealed a multilobulated tumor measuring 4.2 cm in diameter. It was of an erythematous violaceous color and had a hard consistency without becoming stony (falling short of the expected hardness for classic pilomatricoma).

Given the clinical appearance and rapid growth of the tumor, dermatofibrosarcoma, melanoma, and cutaneous metastasis were ruled out. Histology was conclusive and revealed tumor proliferation compatible with pilomatricoma, although there were some uncommon findings for this type of tumor, summarized as a high proliferation index (Figure 2), with: a) abundant matrix cells; b) fewer shadow cells than usual; c) a large number of mitotic cells; and d) elevated tumor proliferation (Ki-67 immunostaining). Histopathology confirmed the benign nature of the tumor and it was completely excised using a bilateral advancement flap H-plasty (Figure 3).

Clinical forms of pilomatricoma include common nodules or small tumors of a hard consistency and are classified according to clinical-pathological features that some authors think correspond to early and late stages in tumor development.¹⁻³ As a result, there is some controversy over whether the same tumor can present different forms or whether each clinical form is truly different even though all cases share a similar origin. To date, the following clinical forms of pilomatricoma have been described:



Figure 1. Large multilobulated tumor proliferation (4.2 cm in diameter) of erythematous-violaceous color and pronounced rapid tumor growth.

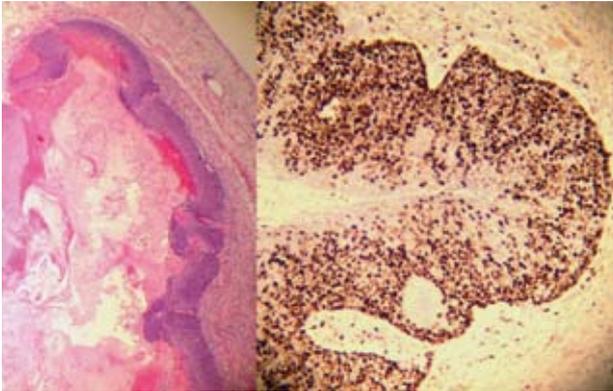


Figure 2. Unusually low number of shadow cells in the panoramic view. Intensely positive Ki-67 immunostaining confirming the proliferative nature of the lesions. (Hematoxylin-eosin, $\times 40$ and Ki-67 $\times 20$)



Figure 3. Removal of the tumor and closure with a bilateral advancement flap H-plasty.

1. Anetoderma associated with pilomatricoma, as the name indicates, described in most texts as an “empty bag,” which often tends to be of a soft consistency with a hard nodule inside. Anetoderma tends appear overlying or adjacent to pilomatricoma.⁴
2. Perforating pilomatricoma, generally of a nodular morphology; presents an erosive surface. These show transepidermal elimination of tumor material consisting fundamentally of calcium, shadow cells, and keratin.^{5,6}
3. The malignant form or pilomatrix carcinoma, which does not appear to originate from a previous benign tumor.⁷
4. Finally, there is the controversial proliferating pilomatricoma.^{8,9} This has not yet been accepted as an independent clinical form, but is described as pilomatricoma that occurs with rapid growth and an abnormal appearance while presenting proliferative changes in histology including a larger number of matrix cells. The characteristics of this disputed form are set out in the Table. These are precisely the changes that occurred in our case and we agree with the above sources that proliferating pilomatricoma presents sufficiently distinct clinical and histological characteristics to warrant classification as an independent entity.

We chose to close the defect using a bilateral advancement flap given our own experience points to the suitability of this type of flap for the affected area. The bilateral advancement flap forms part of a group of sliding flaps that move along the long axis of the plasty.¹⁰ These are mainly used in defects created on the trunk and on the frontal, temporal, maxillary, and labial areas. In

Table 1. Characteristics of Proliferating Pilomatricoma According to Kaddu et al⁸

More common in older patients
Large lesion size
More matrix cells than shadow cells
Greater number of mitosis

fact, the H-flap,¹¹ or double pedicle, is a single or double pedicle plasty that makes use of existing skin creases or wrinkles when used in the frontal area. The technique is implemented by creating a square or round defect, with 2 parallel lines extending from the edges. The distal aspects tend to require the corresponding Burow triangles.

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Conflicts of Interest

The authors declare no conflicts of interest.

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Patient With Malformations of the Face, Mouth, and Fingers: Type I Orofaciodigital or Papillon-Léage Psaume Syndrome

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Type I orofacioidigital syndrome (OFDS), or Papillon-Léage Psaume syndrome (Mendelian inheritance in man



Figura 1. Abnormal facies with hypertelorism, a broad nasal root, a midline cleft of the upper lip, tooth loss, and diffuse alopecia.

605041), is characterized by the presence of orofacial and digital malformations.

The patients often develop dermatologic disorders during the early months of life. The dermatologist can play a key role in establishing an early diagnosis. The disease is sporadic in 75% of cases, while the remainder show an X-linked dominant inheritance. It is lethal in males, and is therefore only observed in females. Its incidence is of 1 in 50 000 live births.¹ The gene responsible for the disease is found on the short arm of the X chromosome (Xp22.2-22.3). The gene product of this fragment may play a fundamental role in organogenesis.²

We present the case of a 29-year-old woman, admitted to the nephrology department due to vascular rejection of a kidney transplant. She was seen for severe generalized pruritus and marked skin dryness during the hospital admission. The important findings on physical examination were an abnormal facies with hypertelorism, a broad nasal root, low-set ears, a cleft upper lip, and the absence of several teeth (Figure 1). She also presented brachydactyly of the fingers of both hands and of some of the toes (Figure 2). The hair was thin, dry, and fragile, with areas of lower hair density most evident at the line of implantation on the forehead.

The patient had been born by normal vaginal delivery after an uncomplicated pregnancy. There was no consanguinity between the parents and karyotyping was 46XX normal. She had multiple phenotypic