Calcified lesions in the skin have been noted since ancient times. Galen, before 200 AD, described stones in some tumors, as did Ambroise Paré in 1585. A more precise description was given by Wilckens in 1856. However, the first complete work, based on a series of patients, was published by Malherbe and Chenantais in 1880. They described calcifying epitheliomas, initially thought to be tumors of sebaceous glands. This view was corrected by Malherbe himself in 1905. The term pilomatricoma, to denote origin from hair matrix cells, was suggested by Forbis and Helwig in 1961. This was later corrected to pilomatrixcoma, as more etymologically correct. These tumors have a wide variety of signs, which often causes misdiagnosis. They are not uncommon and account for one in 500 histologic specimens in our laboratory. We have studied 209 cases that were collected during a 20-year period.

PATIENTS AND METHODS

Case histories of patients with pilomatrixcoma who were seen between 1976 and 1996 were examined. The age, site, size, and physical appearance at examination, and preoperative diagnosis were noted. Histologic evidence confirmed the diagnosis. Additional cases were detected by review of hospital pathology reports and subsequent examination of clinical notes. In total, 209 cases were studied, 125 in female patients and 84 in male patients.

RESULTS

Age at examination

The youngest patient was 18 months of age, and the oldest patient was 86 years of age (Fig. 1). The peak age of presentation was 5 to 15 years in female patients and up to 5 years in male patients. There was a second peak in adults between 50 and 65 years of age. The ratio of female to male patients was 1.5:1 overall but was 2.5:1 in patients less than 20 years of age. Most patients (53%) had a tumor for no longer than 6 months. However, the time to diagnosis ranged from 1 week to 30 years with a peak between 7 to 18 months.

Location

The tumors were predominantly on the head (51.5%), with only 9.5% on the scalp itself. None was present on the palms, soles, or genitalia.
5) or showed telangectasia and a rolled edge, mimicking a basal cell carcinoma (Fig. 6). Rapid enlargement of the tumor as a result of internal bleeding was sometimes noted by patients, bringing it to their attention for the first time. Subsequent hemosiderin pigmentation was seen in 25% of cases, sometimes simulating the appearance of a malignant melanoma.

In 4 cases a further vascular variant was seen. The tumor had a red/blue color (Fig. 7) and appeared to float in a sea of fluid. Sometimes the overlying skin was stretched and atrophic, the so-called anetodermic variant (Fig. 8).

Multiple tumors at the same time were seen in 4 patients. An additional patient, who had had 8 pilomatrixomas removed, also had myotonic dystrophy.

Malignant change

Only 1 patient had a malignant pilomatrixoma. He was 79 years of age and had a presumed epidermoid cyst of the scalp. This was excised, and

Size

Size of tumor at examination varied from less than 0.5 cm to 6.0 cm (Fig. 2) with most tumors (37%) between 1.0 and 1.5 cm.

Clinical appearance

Most tumors appeared as a nodule with a semi-transparent epidermis (Fig. 3). In 25% of cases, calcium was visible as yellow/white flecks that showed through the surface. When the tumor was particularly superficial, calcium eroded through the surface and was extruded (Fig. 4). In one pilomatrixoma of the chest wall, calcification showed on chest radiographic examination, giving the false appearance of tuberculosis on the posterior-anterior view. As many as 20% of the pilomatrixomas were deeply subcutaneous and invisible and were detected by palpation of a hard nodule. The tent sign, first described by Graham and Merwin\textsuperscript{8} in 1978 could be helpful in these cases. Stretching the skin over the tumor allowed the multifaceted nature of the mass to become visible.

Occasionally the tumor had a keratotic appearance, resembling a squamous cell carcinoma (Fig. 5) or showed telangectasia and a rolled edge, mimicking a basal cell carcinoma (Fig. 6). Rapid enlargement of the tumor as a result of internal bleeding was sometimes noted by patients, bringing it to their attention for the first time. Subsequent hemosiderin pigmentation was seen in 25% of cases, sometimes simulating the appearance of a malignant melanoma.

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the diagnosis made histologically. The tumor did not behave aggressively and has not recurred 4 years later.

**Diagnostic accuracy**

The correct preoperative diagnosis was made in 54 cases (21%); all but 6 cases were made by a dermatologist. Five cases were correctly diagnosed by surgeons, and 1 case was diagnosed by a general practitioner. The most common misdiagnosis was that of an epidermoid cyst (38% of cases) with a wide range of other possible diagnoses, including malignancy (8%), inclusion dermoid (7%), hemangioma (5%), or lipoma (4%).

**Treatment**

Treatment of pilomatrixcomas was by incision and curettage or by excision. In some cases, removal of redundant atrophic overlying skin was necessary. No recurrent tumors were noted in our series.

**Histology**

The histologic condition is characteristic with transformation of tightly coherent epithelial cells with basophilic cytoplasm and a round vesicular nucleus to ‘shadow cells’ at the center of the tumor. Calcification, ossification, and foreign body granuloma formation were seen.

In those tumors covered by vascular, atrophic skin, the area between the pilomatrixcoma and the overlying epidermis was filled with dilated lym-
matricomas resemble an epidermoid cyst, others can mimic a variety of malignant tumors. More than one half of the tumors were located on the head, but only 9.5% of them originated on phatic vessels lined by attenuated endothelial cells, in association with numerous small blood vessels and a chronic inflammatory cell infiltrate. We named this the lymphangiectatic variant.

**DISCUSSION**

Our study has confirmed the wide range of presentation of pilomatrixomas. Although many pilo-

**Fig. 4.** Extrusion of calcium through the surface.

**Fig. 5.** Appearance of a squamous cell carcinoma.

**Fig. 6.** Appearance of a basal cell carcinoma.

**Fig. 7.** Lymphangiectatic variant.

**Fig. 8.** Anetodermic variant.

matricomas resemble an epidermoid cyst, others can mimic a variety of malignant tumors.

More than one half of the tumors were located on the head, but only 9.5% of them originated on
the scalp, perhaps surprising for a tumor whose origin is from hair follicles. Noguchi et al.\textsuperscript{10} suggested that the distribution of pilomatricoma corresponds to the density of hair follicles at a particular site. The hairy scalp has about one half the density of follicles of the face, which is the most richly supplied area of the whole body. Our findings would support this view.

The association of multiple pilomatricomas with myotonic dystrophy is well recognized.\textsuperscript{11} Our patient with 8 previous tumors had 2 further lesions. Her myotonic dystrophy had developed 4 years earlier. The family history was unknown, so we were unable to determine whether other members of her family were similarly affected.

Malignant transformation of pilomatricoma is rare,\textsuperscript{12} with fewer than 20 cases described in the world literature. These tumors tend to occur in middle-aged or elderly patients rather than the young patients.\textsuperscript{13} Our case confirmed this view, because he was 79 years of age at the time of examination. The histologic evidence typically shows active proliferating basaloid cells with numerous abnormal mitoses and infiltration into underlying structures.\textsuperscript{14} Tissue necrosis also occurs but was not present in our case.

Although most pilomatricomas have a firm calcified nodule, a few of them look vascular with associated skin thinning. Bowers and Millard\textsuperscript{15} describe this variant in which the typical tumor is covered by irregular vascular spaces that contain lymphatic fluid. Small blood vessels are also increased in number, and the overlying dermis and epidermis are atrophied. In our series, 4 such patients were present, 3 of whom were older than 20 years and thus beyond the age of peak presentation. Jones and Tschen\textsuperscript{16} described a similar series with marked anetodermic skin changes occurring predominantly in young women. The dermis was edematous with attenuated collagen and a marked decrease in the number of elastic fibers. They postulated that catabolic enzymes from the tumor cells or associated inflammatory infiltrate might be responsible for this tissue destruction.

Rarely, a pilomatricoma may perforate the epidermis,\textsuperscript{17} but only if it is formed in the more superficial papillary and mid-dermis, enabling a process of transdermal elimination to occur.

Although treatment of pilomatricoma is ideally by incision and curettage, most tumors in our series had been removed by excision. Only 54 of the 209 cases had been correctly diagnosed before operation, resulting in unnecessarily extensive surgery.

Many tumors (51.5\%) occurred on the head and neck, a cosmetically sensitive area. Consideration of this essentially benign tumor, with careful clinical examination, will lead to increased diagnostic accuracy. This is particularly important for young women in whom most of these tumors occur.

REFERENCES
