

# Pilomatrixoma of the Head and Neck in Children

## A Study of 38 Cases and a Review of the Literature

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**Objectives:** To describe the clinical presentations and discuss the guidelines for surgical management of pilomatrixoma involving the head and neck in children.

**Design:** Retrospective study.

**Setting:** A tertiary care center.

**Patients:** Thirty-three patients, with a mean age of 4.5 years, underwent surgical treatment for pilomatrixoma (n = 38) between 1989 and 1997.

**Intervention:** All patients were treated surgically. In 34 cases, a direct approach was used to achieve complete removal of the lesion with (n = 11) or without (n = 23) skin resection. In the remaining 4 cases, an indirect approach via a parotidectomylike incision was used.

**Results:** In 88% of cases, the presenting symptom was a hard, slow-growing, subcutaneous tumor. The lesion was associated with pain and inflammation in 7 cases (18%) and abscess or ulceration in 4 cases (11%). Twenty-nine patients presented with single nodules and 4 presented with multiple occurrences. The lesions were located on the face (cheek, eyelid, or forehead) in 20 cases (53%), on the neck in 8 cases (21%), in the parotid region in 8 cases (21%), and on the scalp in 2 cases (5%).

**Conclusions:** Pilomatrixoma is a rare, benign skin tumor, but practitioners should be aware of its clinical features. Diagnosis is usually easy based on clinical findings, but computed tomographic scan is helpful, especially in cases involving tumors located in the parotid region. Spontaneous regression is never observed. Complete surgical excision, including the overlying skin, is the treatment of choice.

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**I**N 1880, Malherbe and Chenantais<sup>1</sup> described a benign, subcutaneous tumor arising from hair cortex cells. Since then, this uncommon entity has been called *calcifying epithelioma of Malherbe*. In 1922, Dubreuilh and Cazenave<sup>2</sup> described the characteristic histological features, ie, islands of malpighian cells, including shadow cells and giant cells. In 1961, Forbis and Helwig<sup>3</sup> proposed the name *pilomatrixoma*, thus avoiding the word *epithelioma*, which carries the connotation of malignancy.

Pilomatrixoma usually occurs within the first 2 years of life.<sup>4-6</sup> The most frequent anatomical location is the head and neck region.<sup>4</sup> Most cases involve single nodules,<sup>2,7</sup> but multiple occurrences have been reported.<sup>5,8,9</sup> Familial cases have also been observed<sup>7,9,10</sup> in association with disorders such as Gardner syndrome, Steiner disease, and sarcoidosis.<sup>8,9</sup> Typically, pilomatrixomas are benign but have a ten-

dency to recur if not completely resected.<sup>5,6</sup> A few cases of malignant pilomatrixoma<sup>9,11,12</sup> metastasizing to the lung, bone, brain, abdominal organs, skin, and lymph nodes<sup>11</sup> have been described.

The objectives of this retrospective study were to analyze data from a series of 33 children treated for 38 pilomatrixomas of the head and neck and to discuss diagnostic and therapeutic management.

## RESULTS

There were 17 boys (51.5%) and 16 girls (48.5%). The mean age at the time of diagnosis was 4.5 years (range, 9 months to 14 years). Subdivision according to age showed that 27% of the lesions occurred between birth and 2 years, 63% between 2 and 10 years, and 10% at 10 years or older. The tumor was located on the head in 79% of cases and on the neck in 21%. Four children presented with multiple occurrences either concurrently

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## PATIENTS AND METHODS

Between 1989 and 1997, a total of 33 children presenting with 1 or more pilomatrixomas (n = 38) underwent surgery in the Departments of Otolaryngology and Plastic Surgery, La Timone Children's Hospital, Marseille, France. The records of these patients were reviewed to determine sex, age at initial presentation, age at the time of surgical management, location and size of the tumor, clinical presentation, highlights of the surgical procedure, histological findings, and follow-up findings.

(n = 2) or secondary to treatment of the first lesion at a new site (n = 2). The distribution of the pilomatrixomas was as follows:

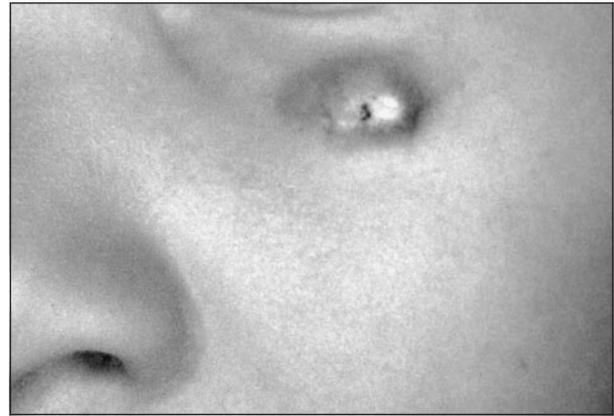
Location	No. (%)
<b>Head (Total)</b>	<b>30 (79)</b>
Cheek	13 (34)
Preauricular region	8 (21)
Eyelids	4 (11)
Forehead	3 (8)
Scalp	2 (5)
<b>Neck (Total)</b>	<b>8 (21)</b>
Lateral area	7 (18)
Posterior area	1 (3)

Two patients had secondary lesions (cheek and lateral area of the neck; preauricular region and scalp and posterior area of the neck), and 2 patients had concurrent lesions (preauricular region and forehead; preauricular region and scalp).

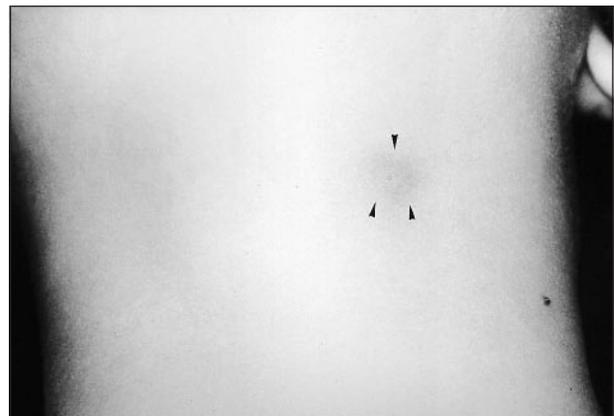
In 88% of cases, the presenting symptom was a stony, hard, subcutaneous tumor sliding freely over the underlying subcutaneous layer (**Figure 1** and **Figure 2**). Most lesions were the size of a pea at the time of consultation and showed a tendency to grow. The mean delay between appearance of the tumor and consultation was 2 months, but 4 patients (12%) reported that their tumors had been present for several years (1-3 years) with no growth (n = 3) or recent changes (n = 1). Pilomatrixomas were associated with pain and inflammation in 7 cases (18%), and the overlying skin showed a tendency to abscess or ulceration in 4 cases (11%). The provisional diagnosis of pilomatrixoma was confirmed in 22 patients. In 3 cases, the provisional diagnosis was angioma owing to the bluish discoloration of the overlying skin. In the remaining 13 cases, no provisional diagnosis was proposed prior to histological study of the surgical specimen.

No concurrent disorders were observed, but 1 female patient's mother and maternal grandmother reportedly had Gardner syndrome, although not associated with pilomatrixoma. In 4 cases, computed tomographic (CT) scan of the parotid region revealed a well-delineated subcutaneous tumor containing microcalcifications (**Figure 3**), and the diagnosis proposed by the radiologist was adenopathy. Sonography was performed in 5 cases (**Figure 4**).

No sign of spontaneous regression was noted in any patient, and surgical excision was performed within a



**Figure 1.** Pilomatrixoma of the left lower eyelid of a 9-month-old boy.

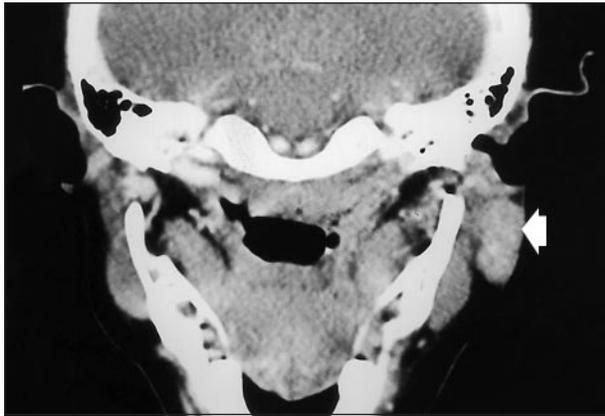


**Figure 2.** Pilomatrixoma (arrowheads) of the left cervical area in a 5-year-old boy.

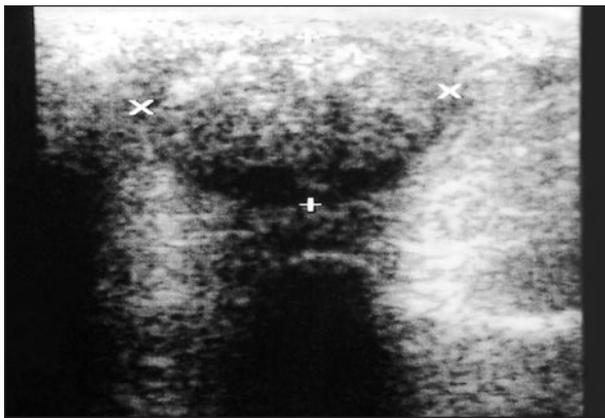
mean delay of 15 months after consultation (range, 1 month to 3 years). In 2 of the 13 cases in which no provisional diagnosis was proposed, medical treatment (cryotherapy and local and systemic antimicrobial therapy) was unsuccessfully attempted. Surgical technique depended on anatomical location. A direct approach starting at the top of the tumor was used without skin resection in 23 cases (61%), a direct approach starting from the tumor with skin resection in 11 cases (29%), and an indirect approach starting at the distance from the tumor via a parotidectomylike incision in 4 cases involving the preauricular region (10%). The size of the surgical specimen ranged from 1 to 2 cm in 36% (n = 14) of cases and was larger than 2 cm in 11% (n = 4) of cases. In the remaining cases, the specimen size was not specified in the histological report.

Histological findings allowed definitive diagnosis in all cases. In most cases, the tumor consisted of islands of cells in circular configuration with enucleated shadow cells in the center and nucleated basophilic cells on the periphery (**Figure 5**). Shadow cells were surrounded by foreign body-type giant cells. In 3 cases (8%), an encapsulated variant was observed. Calcifications were present in 75% of cases.

No recurrence has been observed with a mean follow-up of 3.5 years. However, 2 children developed secondary pilomatrixomas in new locations and underwent surgical excision 5 and 6 years after the first procedure.



**Figure 3.** Coronal computed tomographic image showing an ovoid subcutaneous mass (2.5 × 1.5 cm) in the right parotid region (arrow). Note the intratumoral calcifications.

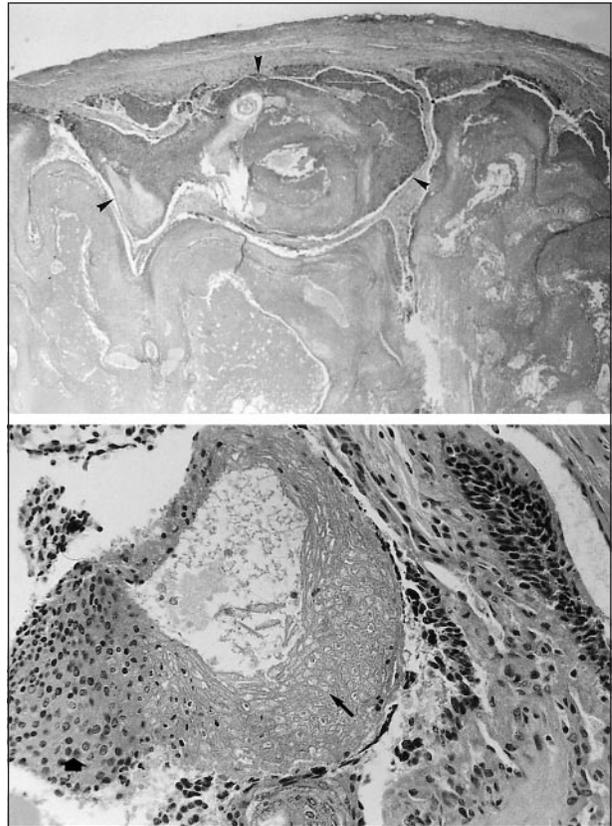


**Figure 4.** Sonogram showing a well-defined hyperechogenic mass (between cursors [x] and plus sign [+]) with a posterior dense acoustic shadow.

## COMMENT

Pilomatrixomas arise from hair cortex cells or the hair follicle of sebaceous glands. The most common anatomical location is the head and neck region. In the meta-analysis reported by Moehlenbeck,<sup>10</sup> head and neck lesions accounted for more than half of the cases (51.8%), followed by lesions on the extremities (37.7%) and trunk (10.5%). No cases of pilomatrixoma have been reported on the palms, soles, or genital region,<sup>3,10,13</sup> to our knowledge, but unusual locations, including the middle ear,<sup>14</sup> have been reported. Based on these anatomical features, several previous authors<sup>14,15</sup> have suggested that the most likely mechanism for development of pilomatrixoma is inclusion of epidermic elements in abnormal locations during embryonic life and subsequent growth after birth. Lever and Griesemer<sup>16</sup> classified pilomatrixoma as a hamartoma, but Dubreuilh and Cazenave<sup>2</sup> proposed 2 alternative hypotheses. The first is that these tumors arise from branchial clefts. This theory seems unlikely because (1) pilomatrixomas can develop at any age and (2) it does not account for lesions on the extremities. The second hypothesis is that pilomatrixomas are of ectodermic origin. This theory is supported by evidence that pilomatrixomas arise from the hair matrix.

Diagnosis of pilomatrixoma in the head and neck can usually be made solely on the basis of clinical features. In



**Figure 5.** Top, Islands of malpighian cells (between arrowheads) including nucleated basophilic cells and enucleated shadow cells (hematoxylin-eosin, original magnification × 100). Bottom, Transition from nucleated basophilic cells (short, wide arrow) to enucleated shadow cells (long, narrow arrow) (hematoxylin-eosin, original magnification × 400).

most cases, tumor diameter ranges from 0.5 to 3 cm, but lesions up to 15 cm have been reported.<sup>17</sup> Most patients are younger than 2 years at the time of discovery,<sup>4,6</sup> and 60% of cases arise before the age of 10 years.<sup>6,13,17</sup> The incidence of pilomatrixoma is much higher in whites than in blacks, and the male-female ratio is 3:2.<sup>4,6,16</sup> Palpation reveals a stony, hard consistency, which is the pathognomonic feature of pilomatrixoma. The tumor slides freely over the underlying layer, while the overlying skin may have a reddish or bluish discoloration, as in 3 cases in our series.<sup>13</sup> The tumor is usually asymptomatic, but some cases have been associated with pain during episodes of inflammation or ulceration. Adenopathy at the time of discovery has never been reported. Pilomatrixomas are usually solitary nodules, but multiple occurrences have been observed in 4% of cases.<sup>3,8,10</sup> Growth is usually slow and benign and occurs over a period of several months or years (3 years in 1 case in our series). However, a malignant variant with distant metastasis to the lung, bone, brain, skin, and abdominal organs has been described.<sup>5,9,11</sup>

According to the literature, the occurrence of multiple or recurring lesions justifies further testing to rule out associated disorders such as Steinert disease, Gardner syndrome, or sarcoidosis.<sup>4-6,9</sup> Some authors consider pilomatrixoma a cutaneous marker of Steinert disease, while others have stated that it is due to a pleiotrophic effect of the gene of the disease.<sup>8</sup> Since other disorders may have similar manifestations, differential diagnosis

may be necessary, depending on the anatomical location, to rule out dermoid cysts in the midline of the neck, parotid tumors in the preauricular region, and adenopathy, calcified hematoma, and lipoma in other sites.<sup>4,5,8,15</sup>

Radiologic imaging is of little diagnostic value for pilomatrixoma. Plain x-ray films show nonspecific calcification,<sup>15</sup> and ultrasonograms demonstrate a well-defined, round, hyperechogenic mass with a posterior dense acoustic shadow. Computed tomography or magnetic resonance imaging (MRI), as described by Fink and Berkowitz,<sup>18</sup> have been used mainly for the relationship between the tumor and the parotid gland. Computed tomography and MRI can be helpful for differentiating preauricular pilomatrixomas from parotid tumors by revealing a sharply demarcated, subcutaneous, opaque lesion that does not enhance after injection of contrast material (CT scan) or small areas of signal dropout that are consistent with the presence of calcifications (MRI scan).

Histologically,<sup>3,16</sup> pilomatrixoma is a deep, subcutaneous tumor occurring between the dermis and hypodermis, with medial displacement of pilosebaceous glands and follicles.<sup>2,15</sup> The tumor is separated from the epidermis by a layer of fibrous tissue. This fibrous layer creates an illusion of adherence to the skin, but there is in fact no connection between the tumor and the epidermis.<sup>2,16</sup> The most common microscopic features are islands of well-organized malpighian cells; maturation is progressive and regular, with no atypical cells. The cells in the islands are arranged in a circular configuration, with nucleated basophilic cells on the periphery and enucleated shadow cells in the center (Figure 5). The islands are associated with a foreign body-type macrophagic reaction and fibrosis. In most cases, the lesions are poorly delineated, but encapsulated forms have also been observed.<sup>6,15</sup> Recurrence is less likely with encapsulated lesions because complete resection is easier. The reported incidence of calcification ranges from 69% to 85%,<sup>10,13</sup> and bone metaplasia is observed in 15% of cases, presumably owing to conversion of fibroblasts into osteoblasts.<sup>3,13</sup> As a result of this process, some lesions are radio-opaque.<sup>4,15</sup> Careful histological study is necessary to rule out invasive forms, which are rare but not impossible. Malignant forms are characterized by a larger epithelial cell component, clusters of undifferentiated basaloid cells, the presence of atypical cells, invasion of blood vessels, and infiltration of capsular tissue.<sup>7,11</sup> At least 2 cases of malignant pilomatrixoma with metastasis have been reported in the literature.<sup>9,11</sup>

Since spontaneous regression is never observed, the treatment of choice is surgical excision. Wide resection margins (1-2 cm) are recommended to minimize the risk of local recurrence.<sup>4,5,9,17</sup> In the literature, it has been reported that incomplete resection has almost consistently been followed by local recurrence.<sup>9</sup> Since the tumor may be tightly adherent to the skin, it may be

necessary to remove the overlying skin (29% of cases in our series). However, the tumor is never adherent to subcutaneous tissue,<sup>2</sup> and separation from underlying tissue is always easy. In the parotid region, the tumor may cover the parotid fascia.

Secondary lesions after surgery are rare, and this risk decreases progressively with age.<sup>6,17</sup> Forbis and Helwig<sup>3</sup> reported an incidence of only 3%. In our series, with a mean follow-up of 3.5 years, there have been no local recurrences, but 2 children developed secondary lesions at a new site and were treated surgically followed by histological observation. McCulloch et al<sup>9</sup> recommend close surveillance based on their experience with 1 patient who, after a series of benign local recurrences, developed a malignant secondary lesion 17 years after the first occurrence.

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