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Characteristics of pilomatrixoma in children: A review of 137 patients

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ABSTRACT

Objective: To describe our institutional experience with pilomatrixoma in the pediatric population, specifically examining its clinical presentation, associated conditions, surgical treatment and future implications.
Methods: This is a retrospective review of a single tertiary care center. Subjects included 137 patients with diagnosis of pilomatrixoma between the years of 2000 and 2013 up to the age of 19. Patient gender, age at excision, number of tumors, tumor size, tumor location, preoperative diagnosis, recurrence, patient race and zip code, along with associated medical conditions were assessed.
Results: There were a total of 174 tumors in 137 patients. The median age at excision was 7.1 years old, with the youngest patient at 6 months and the oldest at 19 years. Head and neck tumors predominated with 70% ($n = 122$) of all pilomatrixomas, followed by the upper extremity with 22% (38). Other locations included the hair-bearing back, chest and lower extremities. A male to female ration of 1:1.2 was observed. Tumor diameter size ranged from 0.2 to 5.2 cm with an average diameter of 1.4 cm. There were no cases of recurrence. Associated diagnoses included Turner syndrome and Sticker syndrome.
Conclusions: Pilomatrixoma, previously thought to be a rare lesion, is one of the most common causes of superficial head and neck masses in children. This study demonstrates clinical presentations that should help guide differential diagnoses. We demonstrate associations that are consistent with the proposed pathophysiology of pilomatrixoma. Surgical excision is curative.

1. Introduction

Pilomatrixoma was first described by Malherbe and Chenantais in 1880 as a "calcifying epithelioma" thought to be of sebaceous gland origin [1]. Forbis and Helwig proposed the term pilomatrixoma in 1961 after elucidating the cortex of the hair follicle as the cell of origin [2]. Thus, this tumor has a predilection for hair-bearing areas of the body. Histochemical and electron microscopy studies support a tumor of hair cell-derived origin. Characteristic features of shadow cells and giant cells have been described [3]. Though under-recognized on cytology in the past, in recent years pilomatrixoma has become a more common diagnosis for cysts in pediatric patients.

The current literature describes pilomatrixoma as a benign tumor typically occurring in pediatric patients, usually within the first decade of life [4]. Adult occurrence of pilomatrixoma has been reported as well, but is often associated with nonspecific malignancies [5]. Thus, a bimodal pattern of occurrence has been suggested, with the first peak being 5–15 years and the second peak occurring at 50–65 years [22]. Pilomatrixoma most commonly occurs in the head and neck. Cases outside of the head and neck region are commonly associated with genetic syndromes and disorders [6–8]. Pilomatrixoma is curative with complete excision. Post-excisional recurrence is rare and most likely due to inadequate surgical excision, and infrequently due to malignancy [20,21,22].

2. Patients and methods

This study was performed with IRB approval. A search of the pathological database of pediatric patients that had undergone mass excision with a pathological diagnosis of pilomatrixoma from Loma Linda University Medical Center (LLUMC) between the years of 2000 and 2013 was done. Patient gender and age at excision, number of lesions, tumor size, tumor location, pathological diagnosis, history of recurrence after 3 months, and patient race were recorded and compared to the current literature. Patient zip code was also noted in order to determine any regional clustering around the large swath of area served by LLUMC. In addition, patient chart review was conducted in search of associated medical conditions.

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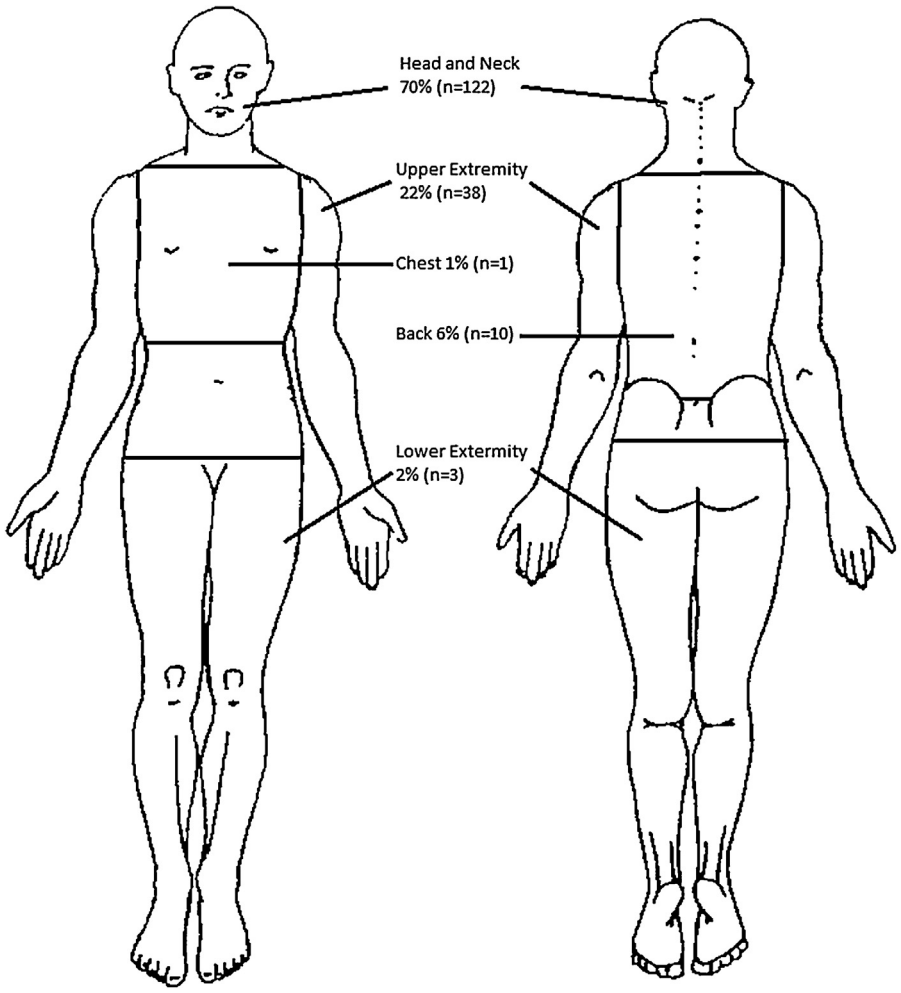

Fig. 1. Incidence of pilomatrixoma throughout the body.

3. Results

Of 137 patients, 55% ($n = 76$) were female and 45% ($n = 61$) were male. The median age at excision was 7.1 years old, with the youngest patient being 6 months old and the oldest, 19 years old. The most frequently encountered age was 5 years old ($n = 17$). Tumor diameter ranged from 0.2 to 5.2 cm with an average diameter of 1.4 cm. 174 total tumors were excised. Among the 137 patients, multiple tumors were found in 14.6% of patients ($n = 20$). 8% of patients ($n = 11$) had multiple lesions involving different body regions. One patient exhibited five separate lesions. 7 patients (5%) of patients had history of previous pilomatrixomas. Correct preoperative diagnosis of "pilomatrixoma" was given in 38% of patients ($n = 52$). The majority of the patients, 53% ($n = 66$) had nonspecific preoperative diagnoses of "mass", "nodule" and "lesion". The remainder of the various preoperative diagnoses included "lymph node", "foreign body" and "epidermoid inclusion cyst". In all cases, the surgical treatment was complete excision. There were no cases of recurrence.

70% of tumors were found in the head and neck, 22% of tumors were found in the upper extremity, 6% of tumors were found in the back, 3 tumors were in the lower extremity, and 1 was in the chest (Fig. 1). The pilomatrixomas found in the head and neck region were further divided into subunits (Fig. 2). 32% were found in the cheek, 20% in the neck, 20% were peri-auricular and 14% were peri-orbital. The remaining subunits including scalp, perioral, nose and

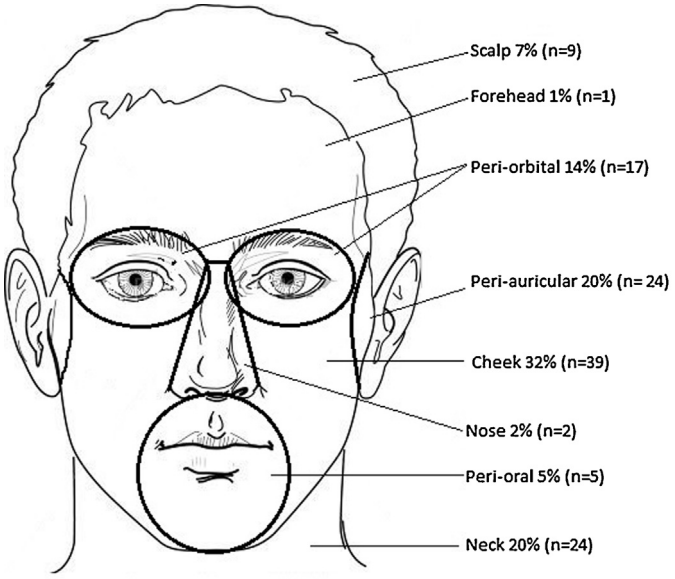

Fig. 2. Pilomatrixoma occurrence in the head and neck.