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Review Article

Review of pediatric head and neck pilomatrixoma

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ABSTRACT

Introduction: Pilomatrixoma is a benign skin tumor, which is commonly found in the head and neck region. It usually presents as an isolated lesion and rarely undergoes malignant transformation. It is not uncommon for these tumors to be misdiagnosed.
Objective: To review the clinical characteristics of head and neck pilomatrixomas in the pediatric population.
Methods: A systematic review was completed by searching ten databases to identify studies reporting findings on pilomatrixoma in the pediatric population. Eligible articles were independently assessed for quality by two authors.
Results: A total of 17 studies met the inclusion criteria totaling 318 pediatric patients. The age of diagnosis ranged from 3 months to 17 years of age. The female to male ratio was 1.65:1. In 14 articles, in which pilomatrixoma was located in the head and neck region, 76 (25%) lesions were found in the neck while 229 (75%) were present in the head region. Three articles exclusively described ophthalmologic pilomatrixoma. Even though pilomatrixoma presents with classical features, the clinical diagnostic accuracy when confronting this lesion averaged 43%. The definitive treatment was surgical excision with a very low recurrence rate.
Conclusion: Head and neck pilomatrixoma in the pediatric population has a typical presentation with a low clinical diagnostic accuracy. Awareness of this lesion and its clinical appearance can improve its diagnosis. We hereby suggest a management algorithm for suspected pilomatrixoma.

Contents

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1. Introduction

Pilomatrixoma is a neoplasm of the hair follicle matrix cells. Malherbe and Chenantais initially described it in 1880 as a calcifying epithelioma, believing it to originate from sebaceous glands. In 1961, Forbis and Helwig proposed the term pilomatrixoma revealing that the cortex of the follicle was indeed the source of origin [1]. Moehlenbeck performed a statistical study of pilomatrixoma cases in 1973. Histological evaluation of 140,000 skin tumor samples at their department of dermatology yielded that pilomatrixoma represented 0.12% of all the samples analyzed [2].

It is most commonly observed in the pediatric population and in the head and neck region. In a clinical review of 209 pilomatrixomas, two peaks of clinical presentation were observed, one important peak occurring from 0 to 20 years of age, and a second less significant peak between 50 and 65 years of age [3]. It has a mean age of presentation of 4.5 years, with 90% of the patients being 10 years old or younger [4].

Clinically, it presents as a firm, stone-like, subcutaneous tumor, which is well demarcated and slides freely over the subcutaneous layer [4]. They can present with overlying normal skin or with a reddish or bluish tint [5]. They usually present as a solitary lesion and can reach up to 3 cm in size or even larger [6]. The diagnosis may be reached by physical examination, imaging and cytology; however, there is a large number of misdiagnosis. This could be attributed to a possible lack of awareness of this tumor by clinicians with proposed diagnoses ranging from benign to malignant skin lesions [7–9]. The definite diagnosis is based on the histological findings, which consist of a well-delineated lesion, originating from the dermis and extending into the subcutaneous fat. They classically consist of islands of epithelial cells containing basophilic cells, ghost cells, and occasionally, foreign body giant cells and calcifications [10].

The purpose of the current systematic review was to collect and analyze all studies published on the diagnosis of children with head and neck pilomatrixoma. This was completed in order to evaluate the epidemiology and clinical characteristics of this distinctive tumor in the pediatric population, and secondly, to increase awareness and decrease difficulties in the diagnosis of this lesion.

2. Methods

2.1. Search strategy

We identified studies by searching electronic databases. The search was performed on February 2015 using multiple databases which included: Africa-Wide Information, AMED (from 1985), Biosis (Previews 1969–2015 Week 12, Previews Archive 1926–1968), Cochrane, Embase (from 1947), Global Health (Global Health 1973–2015 Week 6, Global Health Archive 1910–1972), LILACs, Medline (from 1946), PubMed and Web of science.

The following predefined list of search terms and medical subject headings were used: pilomatrixoma in children (benign lesion) in the head and neck only. The variation of terms used for pilomatrixoma were "epithelioma calcificans of Malherbe", "malherbe calcifying epithelioma" and "(hair follicl* or trichoepithelioma) adjacent to matrix*". The variation of terms used for children were one of "newborn*", "new-born*", "neonat*", "neonat*", "infan*", "child*", "adolesc*", "paediatr*", "pediatr*", "bab*", "toddler*", "kid", "kids", "boy*", "girl*", "juvenile*", "teen*", "youth*", "pubescen*", "parent*", "mother*", "father*", "mom*", "dad*". The search was not restricted to any particular language. Using this strategy, 596 records were identified. Four additional articles were found through other sources totaling 600 articles, of which 330 duplicates were removed by using EndNote's Author/Title/Year duplicate checker (EndNote × 7), followed by a manual verification of duplicates. A dual review process was applied to review the abstracts of all citations identified in the search. After non-applicable records were removed, potentially relevant articles were included on the basis of the full text article. Disagreements were resolved by consensus between the first two authors. Only studies meeting the eligibility criteria were included in this systematic review.

2.2. Inclusion and exclusion criteria

2.2.1. Inclusion criteria

Studies detailing the findings of pediatric patients (age <18 years) with pilomatrixoma in the head and neck region were included. We also included well-constructed retrospective studies and case reports (n > 5 cases). Non-English language studies were also included.

2.2.2. Exclusion criteria

Studies including head and neck pilomatrixoma and other regions without specifying the findings in the pediatric patients were excluded. Studies including adult (age >18 years) and pediatric patients but which did not specify the findings in the pediatric patients were also excluded. Case reports with less than 5 patients were excluded in order to reduce the likelihood of drawing generalizations from isolated case reports. Letters, commentaries, and abstracts were not eligible for evaluation.

2.3. Data extraction

Extracted data included patient demographics, number of patients, female to male ratio, tumor features, diagnostic characteristics, management and outcomes. We extracted pediatric patient data from studies involving both children and adults when specific details of each patient were available, and those involving head and neck as well as other regions.

3. Results

A total of 270 articles were screened with 114 being excluded due to unmatched key terms. After reviewing the abstracts and adding 26 full text articles with unavailable abstracts, 122 were eligible for full text review. Three articles could not be retrieved from the North American library system and were therefore excluded [11–13]. All together one hundred and five full text articles were excluded. We could not isolate the data of the pediatric patients from those of adults, or head and neck pilomatrixoma from other regions within these articles. At times, even if we were able to isolate the data, it did not meet the criteria for minimum amount of patients (n > 5). We found 17 articles to be eligible [4,5,7,10,14–24] for the systematic review, of which 3 were specifically ophthalmology oriented [5,14,15]. The flow diagram of the search is shown in Fig. 1. Fifteen studies are retrospective cohorts [4,5,7,10,14,15,17–19,21–26], one is a prospective study [16] and one is a case series [20].

3.1. Patient demographics

A final cohort of 318 pediatric patients was evaluated totaling 338 tumors in the head and neck region (Table 1). The number of patients included in each of the articles ranged from 6 to 86 patients. In total, we observed 198 females and 120 males with a female to male ratio of 1.65:1. Fifteen articles gave information on the age of