



Pilomatricoma: Clinical, Dermoscopic Findings and Management in 55 Pediatric Patients and Concise Review of the Literature with Special Emphasis on Dermoscopy

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ABSTRACT Introduction: Pilomatricoma is a benign adnexal dermal or subcutaneous tumor derived from immature hair matrix cells.

Objectives: The aim of our study is to evaluate clinical and dermoscopic features of pilomatricomas, with a specific focus on pediatric lesions, and to provide a concise review of the existing literature.

Methods: A single-center retrospective study was undertaken on 55 patients with a histopathological diagnosis of pilomatricoma referred to the Dermatology Unit, University of Bologna, Bologna, Italy, between 2005 and 2023. Pilomatricomas were retrospectively evaluated relying on clinical and dermoscopic images. A PubMed search was conducted. All the relevant research up to July 31, 2023, was reviewed. We classified the cases as “typical” or “atypical” based on whether they were suspected of being pilomatricomas or not.

Results: A total of 55 children with pilomatricomas were observed and studied. Two patients presented with 2 pilomatricomas, leading to the identification of 58 pilomatricomas. ‘Typical’ pilomatricomas were observed in 79% of cases as nodular and pigmented lesions with one or more colors, ranging from blue-gray to red to yellow/white, evident on clinical examination and even better on dermoscopy. In 21% of cases, pilomatricomas presented in an ‘atypical’ form, which did not allow for a well-founded suspicion, placing them in differential diagnosis with other lesions and therefore requiring histological examination.

Conclusions: According to our case series and systematic review of the literature, clinical appearance and dermoscopy may be sufficient to diagnose or suspect pilomatricoma in around 80% of cases, while histological examination is necessary to confirm the diagnosis in the remaining 20% of cases.

Introduction

Pilomatricoma, also called “calcifying epithelioma of Malherbe” or “pilomatrixoma”, is a benign adnexal dermal or subcutaneous tumor derived from immature hair matrix cells. It was first described in 1880 by Malherbe and Chenantais and, in 1961, Forbis and Helwig coined the term “pilomatrixoma” [1,2].

This tumor typically affects individuals during their first two decades of life. Clinically, it usually manifests as a firm, deep nodule, with a diameter ranging from 3-30 mm, mainly on the upper body.

Although the pediatric onset is the most common, dermoscopic features are primarily reported in adult lesions and only a few cases of pediatric patients have been described in the literature (Table 1) [3-13].

Objectives

The aim of our study is to evaluate both typical and atypical clinical and dermoscopic features of pilomatricomas, with a specific focus on pediatric lesions, and to provide a concise review of the existing literature.

Methods

A single-center retrospective study was conducted on 55 patients with a histopathological diagnosis of pilomatricoma referred to the Pediatric Dermatology Unit, University of Bologna, Bologna, Italy from 2005 to 2023.

Pilomatricomas were retrospectively evaluated by four dermatologists with expertise in dermoscopy and pediatric dermatology, relying on clinical and dermoscopic images only, and not allowed to consult histology and ultrasound investigations.

A pilomatricoma was considered “atypical” if at least 3 out of 4 dermatologists disagreed on the diagnosis of pilomatricoma. Two groups were therefore created: lesions that

were suspected of being pilomatricomas, and lesions that were not. In the latter cases, other differential diagnoses were suggested.

We identified all the studies indexed in PubMed until 31 July 2023. All papers reported in the present study were based on clinical studies involving humans, including case reports, case series and reviews. The search parameters included the terms “pilomatricoma in children”, “dermoscopic findings in pilomatricoma”, “typical dermoscopy of pilomatricoma”, “atypical dermoscopy of pilomatricoma” and “pilomatricoma dermoscopy”. A subsequent review of the respective bibliographies aimed to identify any undetected reports. Apart from two articles in German and French, only papers written in English were considered in the review.

Results

We observed and studied 55 children with pilomatricomas. Results are summarized in Table 2.

Females were involved in 60% of cases. Mean age was 7 years. Two patients presented with 2 pilomatricomas without extracutaneous manifestations, leading to the identification of 58 pilomatricomas in our retrospective analysis.

Thirty-eight of 58 (65%) pilomatricomas were located on the head-neck region, 14 (24%) on the upper limbs and a few cases on the lower limbs, back and abdomen.

Clinically, pilomatricomas typically present as pigmented nodules. Forty-six pilomatricomas in our case series (79%) exhibited single or multiple colors ranging from blue to red or yellow (Figure 1, A, C and E). The nodules were exophytic, ulcerated, or subcutaneous in 17.2% of cases (Figure 1G). The mean diameter of the lesions was 7 mm.

Furthermore, rare clinical types of pilomatricomas characterized by anetodermic or keloid-like appearance were described (3% of cases).

An ultrasound scan was prescribed for 24 children (44%) and the report included the following features: dimensions,

Table 1. Overview of the Literature Review.

Study and Year of Publication	Type of Study	Patients			Cutaneous Area Involved	Typical at Dermoscopy	Atypical at Dermoscopy
		N. of Patients Involved With Dermoscopy Available	Case N.	Age and Sex			
Pedro Zaballos et al, 2008 [3]	Clinical study	10	1	75, F	Arm	Tricolor pathognomonic appearance	
			2	40, M	Arm		
			3	45, M	Arm		
			4	12, F	Face	Tricolor pathognomonic appearance	
			5	36, F	Neck		
			6	52, F	Face	Tricolor pathognomonic appearance	
			7	16, M	Face		
			8	18, F	Arm		
			9	14, F	Face		
			10	60, F	Face		
Hernández-Núñez et al 2014 [4]	Retrospective Study of Pilomatricoma	1	1	Children	Face	Tricolor pathognomonic appearance	
Martínez-Moran et al, 2014 [5]	Case report	1		61, F	Right frontoparietal region	Tricolor pathognomonic appearance	
Ivette Alarcon et al, 2014 [6]	Case report	1		12, M	Left cheek	Tricolor pathognomonic appearance	
Erhan Ayhan et al, 2014 [7]	Case report	3	1	67, F	Right cheek		Molluscum-like appearance
			2	48, M	Left eyebrow	Nodular bicolor appearance	
			3	58, M	Left side of the nose	Nodular bicolor appearance	
Wolff et al, 2014 [8]	Case report	1		4, M	Left cheek		Nodular pilomatricoma with blue-red color mimicking vascular lesion
Chen et al, 2020 [9]	Case report	1		14, M	Left arm	Nodular bicolor appearance	
Neema et al. 2022 [10]	Case report	2	1	34, F	Left cheek	Tricolor pathognomonic appearance	
			2	42, F	Right pinna	Tricolor pathognomonic appearance	

Table1 continues

Table 1. Overview of the Literature Review. (continued)

Study and Year of Publication	Type of Study	Patients			Cutaneous Area Involved	Typical at Dermoscopy	Atypical at Dermoscopy
		N. of Patients Involved With Dermoscopy Available	Case N.	Age and Sex			
Anubha Dev et al, 2022 [11]	Case report	3	1	40, M	Right shoulder		Anetodermic pilomatricoma
			2	42, M	Left mammary area		Anetodermic pilomatricoma
			3	70, F	Left forehead region		Anetodermic pilomatricoma
Fink et al, 2017 [12]	Case report	1	1	3, M	Right zygomatic arch		Molluscum-like appearance
Huet et al, 2018 [13]	Case report	1	1	10, M	Left temporal region	Tricolor pathognomonic appearance	

margins, depth, both hypoechogenicity and hyperechogenicity, hypochoic rim, shadow cone, calcifications, non-homogeneous ecostructure. In 5 cases (21%) lesions had regular margins. The shadow cone was documented in 9 cases (37%) and in 10 cases (42%) there was calcification. Among the lesions presenting calcification, 8 (80%) were coarse formations, while 2 (20%) had a dotted appearance.

Concerning ultrasound examination, in 15 patients (62%), it was able to confirm the diagnosis of pilomatricoma. In 8 (33%) it did not contradict the clinical hypothesis of pilomatricoma, while in 2 (8%) it was inconclusive.

In 36 of the 58 examined cases (62%), pilomatricomas presented as pigmented exophytic nodules, ulcerated or not, red and/or blue-gray in color, with or without visible vessels on the surface, classifiable as pigmented nodular type (Figure 1, A and B). In 24 cases (41%), mainly located on head-neck region, pilomatricomas were characterized by blue-yellow-reddish homogeneous areas, representing a pathognomonic tricolor nodular type (Figure 1, C-F). If only 2 colors were present, lesions were still suspected to be pilomatricomas (bicolor type lesions). In 10 cases (17%), pilomatricomas were classified as deep because of the presence of a pseudonetwork and a slight white-yellowish perifollicular discoloration associated with the underlying calcifications (Figure 1, G and H). In such cases, a hard nodule on palpation may also suggest a calcified cyst as a differential diagnosis.

Diagnosis of pilomatricoma was suspected in 79% of cases based on clinical and dermoscopic evaluation.

The clinical and dermoscopic presentation was atypical or not suggestive in 12 of 58 cases (21%).

Among them, 4 pilomatricomas (33%) presented as small nodules with a central white-yellowish component, surrounded either by an erythematous (red) halo or by crown vessels. In these cases, a dermoscopic diagnosis of molluscum contagiosum was proposed (Figure 2, A and B). In 4 cases (33%), pilomatricomas presented as subcutaneous nodules with a red or blue background and arborizing-like or reticular pattern linear-irregular flat vessels, mimicking calcified vascular lesions (Figure 2, C and D). Two pilomatricomas (17%) resembled melanocytic lesions showing a brown pseudonetwork or homogeneous blue pattern at dermoscopy (Figure 2, E and F). In 2 cases (17%) the clinical presentation was keloid-like. Furthermore, biopsy of a lesion was performed to exclude a possible malignancy such as dermatofibrosarcoma protuberans (Figure 2, G and H).

Histological examination revealed a relatively well-circumscribed, dermal or dermal-subcutaneous, multilobulated tumor, surrounded by a variable connective tissue stroma. The tumor consisted of islands of cells in a circular distribution with anucleated shadow cells in the center and basophilic cells in the periphery.

In addition, certain distinctive histological characteristics may be associated with the particular clinical-dermoscopic variant. Tricolor pilomatricomas, for example, showed calcifications or melanin pigment in the lobules (Figure 3A). Dilated blood vessels overlying the tumor were observed in the vascular type (Figure 3B). Keloid-like pilomatricomas, on the other hand, featured a loss of collagen and elastic fibers in the superficial dermis overlying the lesion. Dermoscopic-histopathological correlation showed that bluish areas were associated with the presence of melanin pigment, white areas

Table 2. Epidemiological Data, Anatomical Site, Clinical and Dermoscopic Findings of Pilomatricomas.

General Data	
Number of patients	55
Number of pilomatricomas	58
of which are atypical	12 (21%)
Epidemiological Findings	
Mean age at diagnosis (years)	7.2
Male gender, N (%)	22 (40%)
Female gender, N (%)	33 (60%)
Anatomical Site	
Face	35 (60.3%)
Upper extremities	14 (24%)
Lower limbs	2 (3.4%)
Back	3 (5.2%)
Neck	3 (5.2%)
Abdomen	1 (1.7%)
Patients With Multiple Lesions	2 (3.6%)
Dermoscopic Features	
Yellow-white structureless areas	29 (50%)
Perilesional Yellow area	2 (3.5%)
Central yellow area	2 (3.5%)
Reddish homogeneous areas	42 (72.4%)
Linear irregular unfocused vessels	36 (62%)
Dotted vessels	4 (6.9%)
Structureless blue-gray areas	27 (46.5%)
Ulceration	7 (12%)
Clinical-dermoscopic Findings of Typical or Suspected Pilomatricomas	
Pigmented bicolor nodular type	12 (20.7%)
Pigmented tricolor nodular type	24 (41.4%)
Deep pilomatricomas with pseudonetwork and follicular openings	10 (17.1%)
Clinical-Dermoscopic Findings of Atypical Pilomatricomas	
Molluscum-like pilomatricomas	4 (6.9%)
Vascular-like pilomatricomas	4 (6.9%)
Melanocytic-like pilomatricomas	2 (3.5%)
Keloid-like pilomatricomas	2 (3.5%)

with calcification, red areas with dilated blood vessels and white streaks corresponded to fibrosis on histopathology.

Conclusions

According to our study and the review of the literature, pilomatricoma is more common in females and it is mainly

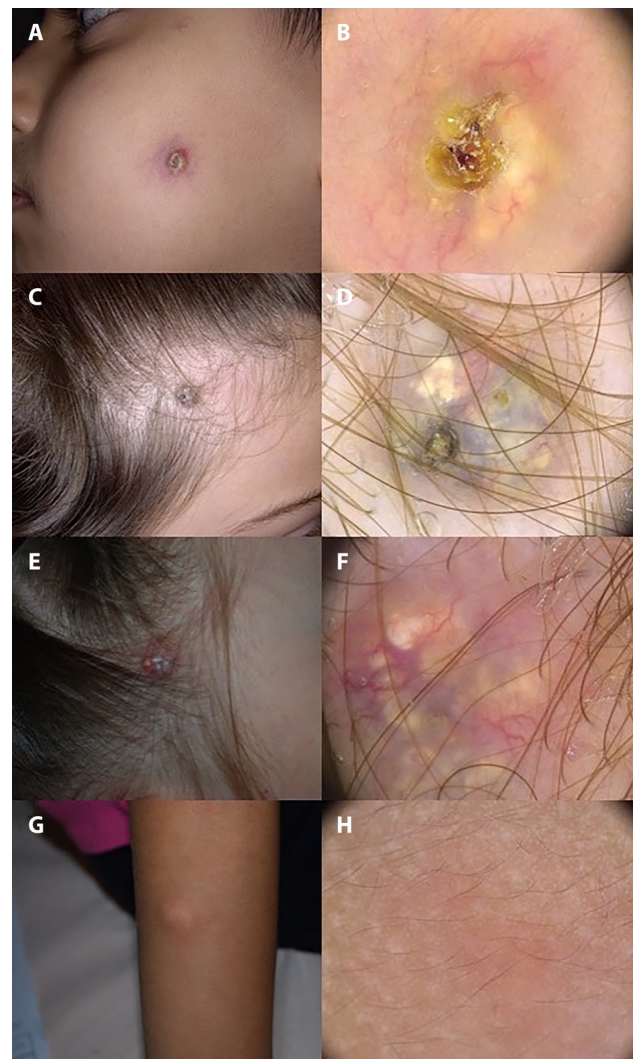


Figure 1. (A) Ulcerated nodule with erythematous halo on the face of a 10-year-old child. (B) Bicolor yellow-white structures with unfocused linear-irregular vessels around the lesion. (C-F) Exophytic red, blue-gray, whitish nodule localized on the head and neck respectively; dermoscopy shows a pathognomonic 3 colors appearance characterized by blue-yellow-reddish homogeneous areas plus linear irregular vessels. (G,H) Deep variant of pilomatricoma: subcutaneous nodule surrounding by normal skin with mild skin changes at clinic and dermoscopy.

localized on the head-neck region [14]. Multiple pilomatricomas may be associated with myotonic dystrophy, familial adenomatous polyposis-related syndromes (including Gardner syndrome), Turner syndrome, or Rubinstein-Taybi syndrome [15,16]. Ciriacks Ket al found that the presence of six or more pilomatricomas is highly suggestive for an underlying syndrome (>95% specificity), so these patients should undergo additional screening [15]. In our retrospective analysis, two children presented with multiple pilomatricomas but further investigations were negative.

Pilomatricomas may present with a varied morphology, therefore, making preoperative diagnosis challenging. The literature highlights an exophytic pigmented lesion as the



Figure 2. (A,B) Shiny papulo-nodular lesion localized on the face of a 9-year-old child showing homogeneous yellow-white central area with crown vessels at dermoscopy. (C,D) Subcutaneous red nodule on the malar region of a 12-year-old child characterized by large linear-irregular vessels on yellow-whitish background at dermoscopy. (E,F) Flat blue lesion localized on the neck of a 11-year-old child, dermoscopy shows light blue homogeneous areas. (G,H) Plaque irregular in shape 1.5cm diameter nodule, localized on the back of a 8-year-old child, dermoscopy shows an unspecific red homogeneous pattern with unfocused vessels.

typical and suggestive presentation of pilomatricoma in the pediatric population. Such lesions can be ulcerated or non-ulcerated and may exhibit variable pigmentation with combinations of blue-gray, red, and white/yellow colors [4,6-8,10,12-14]. Additionally, atypical pilomatricomas with an anetodermic appearance have also been reported [5,9,11]. In the literature, ultrasound was used to confirm, support or exclude the diagnosis of pilomatricoma [18,19]. Once the diagnosis has been confirmed, surgical excision may be considered for aesthetic and functional reasons.

Dermoscopy may be a useful tool in order to improve the detection of pilomatricomas. In the largest case series described by Zaballos et al, the use of dermoscopy increased the diagnostic sensitivity for pilomatricoma from 50% to 90% [3]. On the other hand, in our retrospective analysis, clinical and dermoscopic features were suspicious or suggestive of pilomatricoma in only 79% of children, while in 21% of cases, they were insufficient to make the diagnosis and histology was required. Zaballos et al reported the presence of multiple irregular whitish structures and streaks on dermoscopy in 90% of cases [3]. In contrast, in our retrospective study, yellow/whitish areas were found only in 50% of pilomatricomas. These differences in sensitivity and dermoscopic findings may be related to the larger and different sample size (55 pediatric patients included in our analysis versus only 3 children described by Zaballos et al).

The systematic review of the literature highlighted that both cases of typical and atypical pilomatricomas have been reported.

The tricolor appearance could be considered as a dermoscopic clue to make the diagnosis. In this regard, several authors reported typical pilomatricomas characterized by a bi- or tricolor nodular aspect at dermoscopy [3-6,9,10,13].

In contrast, dermoscopy of atypical pilomatricoma has rarely been reported in the literature [7,8,11,13-15]. Several authors have described nodules characterized by yellowish lobules on an erythematous background, surrounded by crown-like branching vessels features that can also be found in molluscum contagiosum, which must be considered as a differential diagnosis [7,13].

Wolff et al reported a 4-year-old boy with a red-blue nodule, 0.5 cm in diameter, characterized by a homogeneous blue-red color and linear white structures on non-polarized dermoscopy. Histology was required to exclude a calcified hemangioma. In addition, Dev et al reported three cases of anetodermic pilomatricoma, which is extremely rare and presents clinically with a scar-like appearance [11,15]. Dermoscopy is inconclusive in these cases and dermoscopic-histopathological correlation is essential to differentiate between dermatofibroma and dermatofibrosarcoma protuberans [15].

Clinical and dermoscopic findings of pilomatricoma in pediatric age are poorly reported in the literature.

Our large case series and systematic review of the literature indicate that in approximately 80% of cases dermoscopy may be sufficient to diagnose or suspect pilomatricoma, whereas in the remaining 20% of cases histological examination is necessary to confirm the diagnosis and exclude malignancy of the lesion.

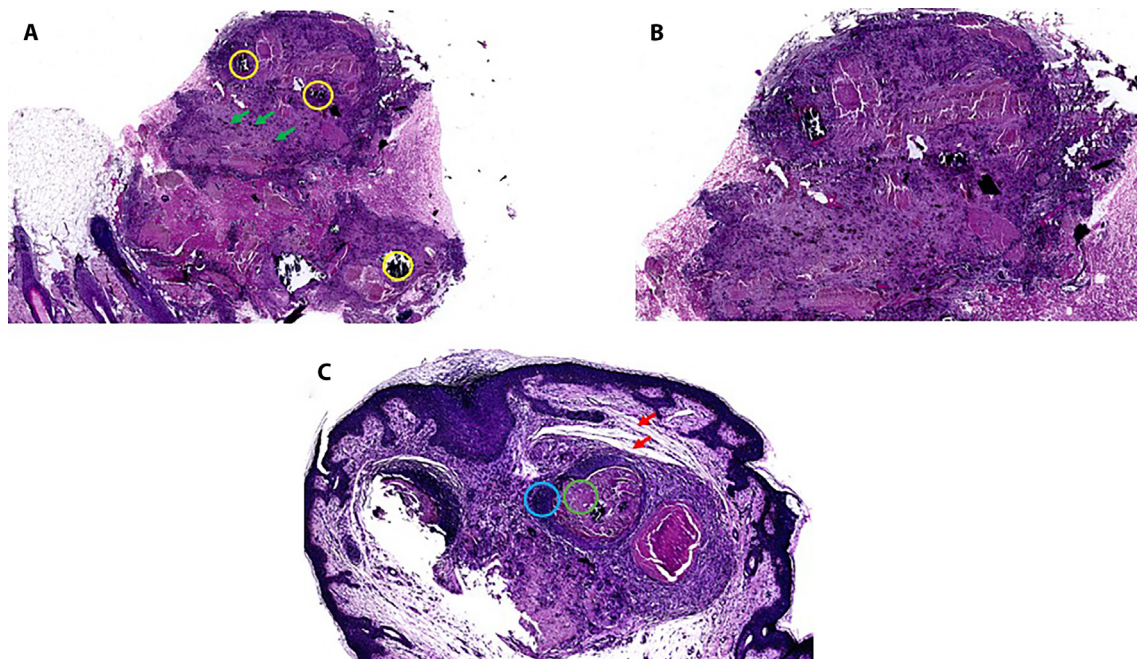


Figure 3. (A) Tricolor type pilomatricoma: peripheral aggregates of basaloid cells and central large masses of eosinophilic cornified material containing shadow cells. Calcification (yellow circles) and melanin pigment (green arrows) are present into the lobules (H&E, $\times 60$). (B) Tricolor type pilomatricoma (H&E, $\times 90$). (C) Vascular type of pilomatricoma: non-capsulated tumor proliferation in the central area of the dermis, with basaloid cells (blue circle) with an abrupt transition to shadow cells (green circle), dilated vessels between the tumor and the epidermis (red arrows) (H&E, $\times 90$).

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