PILOMATRICOMA – CLINICAL, DERMATOPATHOLOGICAL AND DERMATOSCOPIC REVIEW

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Summary

Pilomatricoma (pilomatrixoma) is a tumor that originates from the matrix of the hair follicle. It is the most common follicular tumor, and also the most frequent skin tumor of the child. The incidence of pilomatricoma shows two main peaks: in the first two decades of life and in the sixth decade.

Clinically it presents as a firm or hardened nodule with dimensions of about 1 cm with slow evolution usually located on the face and sometimes on the upper limbs. The clinical signs encountered in pilomatricom are: the tent sign, „teeter-tother sign", skin crease sign that allows the diagnosis in 50% of cases. Histologically, the presence of „shadow cells" that preserve the structure of the cell membrane and traces of the nuclei is characteristic. Dermatoscopic white-yellow structures, white streaks and vascular structures appear.

The evolution is benign, and the treatment is surgical.

***Keywords**: pilomatricoma, pilomatrixoma, hair follicle, child, shadow cells, white streaks.*

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Malherbe calcifying pilomatricoma or epithelioma is a benign skin tumor that originates from the pillar matrix, accounting for 0.1% of skin tumors. Often confused with other benign or malignant skin tumors, pilomatricoma is one of the most frequent follicular tumors. Malherbe and Chenantais described it in 1880 as „epithelioma calcifying ou momifie des glandes sebacees" and hypothesized that the lesion came from a sebaceous gland and therefore called the tumor calcifying epithelioma of the sebaceous glands [1].

Forbis and Helwig in 1961 showing the origin of the tumor in the pillar matrix call it pilomatrixoma [2]. Currently this has been corrected and the term pilomatricoma is used, which etiologically seems more correct, given by Julian in 1998 [3].

Pilomatricoma is a tumor that occurs more frequently in young people up to 20 years of age (60% of cases), 40% of cases occurring in children up to 10 years. However, it is reported that pilomatricoma can occur at any age, with 15% of cases occurring in the sixth decade of life. It most often affects women (sex ratio 1/2 to 1/3) before the age of 20.

Clinically, the pilomatricoma presents as a single node, rarely multiple, firm or hard, deeply dermal or subcutaneous, well circumscribed more or less highlighted by the surface of the skin, with slow asymptomatic growth, rarely painful. It adheres to the superficial planes but

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not to the deep one, having an irregular and bumpy surface, giving the sensation at palpation of facets and angles. The diameter varies between 3 and 30 mm. The color is blue, reddish, white or normal skin. The hard and polygonal character of the tumor is evocative of the diagnosis of pilomatricoma. The location of the tumor is predominantly on the face (cheeks, temples, forehead, preauricular region) and neck, sometimes the upper limbs, but rarely on the trunk and lower limbs. It has rarely been reported to be preceded by trauma to the site [8].

A number of clinical signs for the diagnosis of pilomatricoma are cited [5]:

- The tent sign (Graham and Merwin), when stretching the slightly anetodermal skin overlying the tumor is felt through the atrophic dermis, the bumpy character and the multiple faces and angles of the pilomatricoma can be visualized;
- „Teeter-totter sign", applying pressure to one edge of the tumor causes the protrusion of the opposite edge (rocker);
- Finally, the skin crease sign consists of pinching the lesion with the nails of the two thumbs and highlighting a longitudinal skin crease.

Several clinical forms of pilomatricoma have been described, such as perforating, ulcerating, anetodermal, bullous or pigmented. Single pilomatricoma does not require special investigation, but family forms or forms with multiple lesions may be dermatological markers of conditions such as myotonic dystrophy, Gardner syndrome, or Rubinstein-Tayb syndrome [4].

The clinical diagnosis of pilomatricoma can be evoked only in 50% of cases, the differential diagnosis being made with numerous tumors, the main ones being the epidermoid cyst and the dermoid cyst. The epidermoid cyst differs from pilomatricoma in its pasty consistency and the presence of a small apical orifice through which a yellowish-white substance with a characteristic rancid odor can be seen. Histologically, the epidermoid cyst is bordered by a keratinized malpighian sheath. Dermoid cyst is more common in children and adheres to superficial planes. Histologically it is a cyst with an epidermoid epithelial wall associated with papillary and mesenchymal structures. Other diagnoses that can be discussed in children are juvenile xanthogranuloma, idiopathic facial aseptic granuloma, Spitz's nevus, hemangioma, lipoma, and in the elderly with basal cell or squamous cell carcinoma, Merkel tumor.

Confirmation of the diagnosis of pilomatricoma is histological. It shows a well-defined, multilobular dermal tumor formed on the periphery of basophilic cell islands, and in the center of wide islands of mummified cells called shadow cells whose structure preserves the membrane silhouette and spectral traces of nuclei (these mummified cells) corresponding to a maturation incomplete abortion pillars. Foreign body giant cell are constantly observed between the cell islands. Calcification of mummified cells is common but inconsistent. Transepidermal release of shadow cells can sometimes be seen [7].

Dermatoscopy first used by Pedro Zaballos in 2008 highlighted the following features of pilomatricoma [6]:

- multiple white-yellow structures, well delimited with irregular shape and distribution (80% of cases) and white streaks (70% of cases) corresponding to calcifications and shadow cell areas;
- vascular structures present in 100% of cases most often in the form of homogeneous red areas due to the presence of numerous vessels in the papillary dermis and hemorrhages (90%), hairpin vessels (70%), linear vessels irregular (60%), dotted vessels (40%) and atypical vessels;
- ulcerations caused by an external trauma or perforation;
- small gray-blue areas without structure due to intracellular melanin deposits or the presence of melanophages or siderophages in the inflammatory infiltrate.

The presence of these multiple colors in dermatoscopy is sometimes described as the „rainbow pattern".

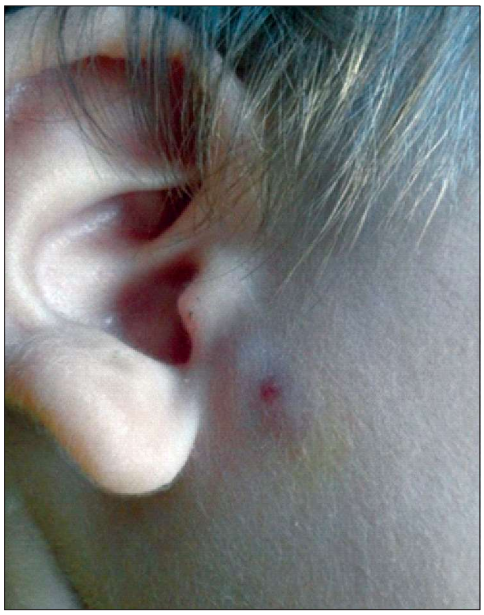

Figure 1. Pilomatricoma – 3 year old pacient

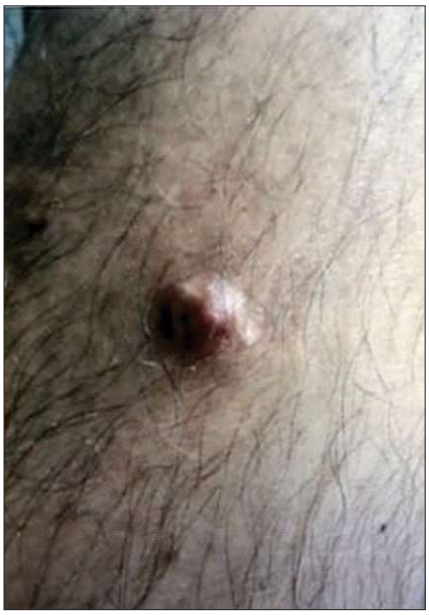

Figure 2. Pilomatricoma – 43 year old pacient

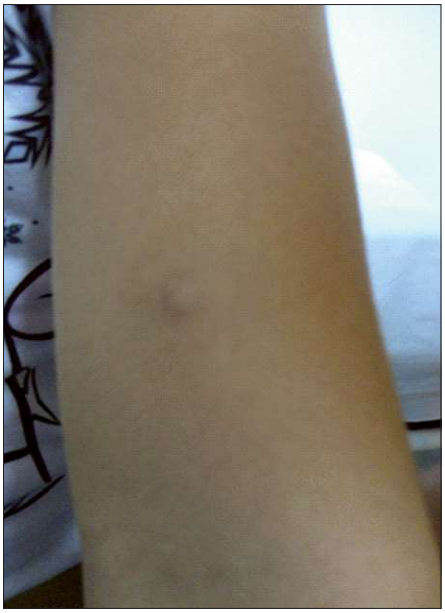

Figure 3. Pilomatricoma – 11 year old pacient

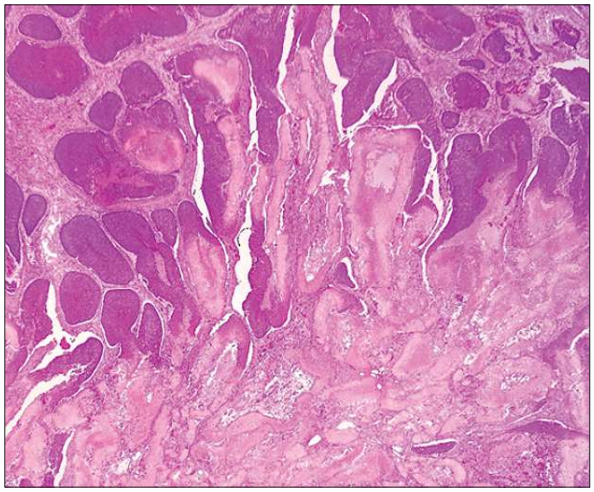

Figure 4. Biphasic tumor proliferation with basaloid cell islands and lobules progressing to nucleus-free eosinophilic cell islands (shadow cells)

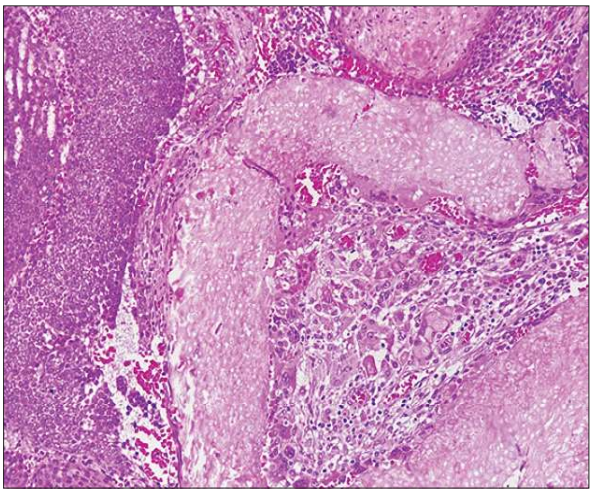

Figure 5. The transition from the basaloid component, with immature cells and the eosinophilic component, made up of shadow cell ranges. Infiltrated with giant multinucleated histiocytes that accompany the keratinized component, matrix

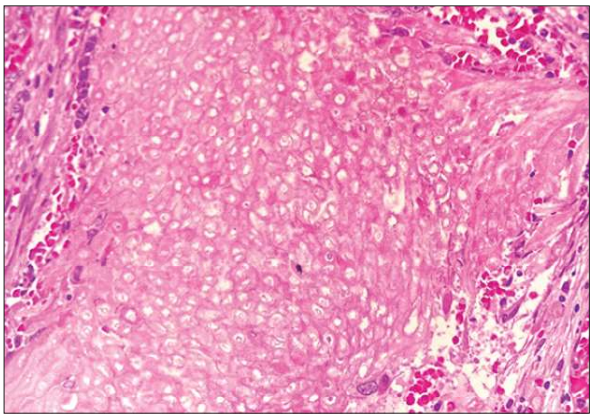

Figure 6. Shadow cell islands

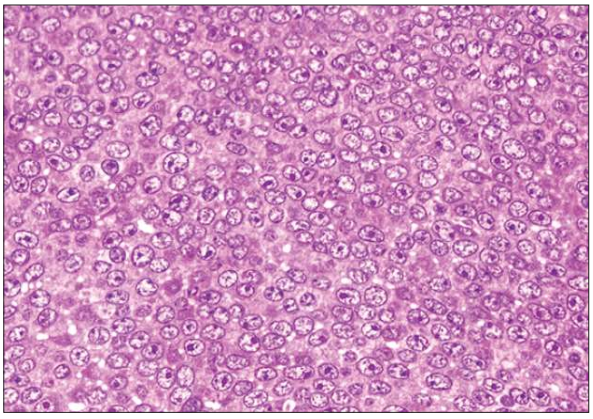

Figure 7. Basaloid tumor component: immature cells, quantitatively reduced cytoplasm, round nuclei, vesicles, prominent nucleoli, scattered mitosis (mimics the bulbar portion of the follicular epithelium)

Doppler ultrasound and magnetic resonance imaging show a well-defined subcutaneous „cystic" lesion with a thickened vascularized wall with small calcifications. The contents are inhomogeneous, with no liquid level or partition walls.

It should be noted that pilomatricoma is a benign tumor with a slow evolution in the majority of cases. Malignant transformation is rarely described in elderly subjects. Surgical excision is required if the pilomatricoma is a tumor that never regresses spontaneously and is not followed by recurrence.

In conclusion, the diagnosis of pilomatricoma should be made in the presence of a hard tumor without a tendency to regress in a child.

Bibliography

1. Malherbe A, Chenantais J. Note sur l'epithelioma calcifie des glandes sebaces. Progres Medical. 1880;8:826–828. [Google Scholar]
2. Forbis R, Jr, Helwig EB. Pilomatrixoma (calcifying epithelioma) Arch Dermatol. 1961 Apr;83((4)):606–18. – PubMed
3. Julian CG, Bowers PW. A clinical review of 209 pilomatricomas. *J Am Acad Dermatol*. 1998;39(2 Part 1):191–195. [] [
4. Ciriacks K, Knabel D, Waite MB.Pediatr Dermatol. 2020 Jan;37(1):9-17. doi: 10.1111/pde.13947. Epub 2019 Oct 16
5. Huet P, Barnéon G, Cribier B.Ann Dermatol Venereol. 2018 Aug-Sep;145(8-9):539-543. doi: 10.1016/j.annder.2018.05.007. Epub 2018 Jul 17.
6. Zaballos P, Llambrich A, Puig S, Malvehy J.Dermatology. 2008;217(3):225-30. doi: 10.1159/000148248. Epub 2008 Jul 25
7. Huet P, Barnéon G, Cribier B.Ann Dermatol Venereol. 2018 Aug-Sep;145(8-9):539-543. doi: 10.1016/j.annder.2018.05.007. Epub 2018 Jul 17.
8. Yalcin NG, Mann N.ANZ J Surg. 2019 Sep;89(9):E390-E391. doi: 10.1111/ans.14566. Epub 2018 May 15.

Conflict of interest
NONE DECLARED

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