

Available online at www.sciencedirect.com

journal homepage: www.elsevier.com/locate/radcr

Case Report

Pilomatrixoma of the right thigh: Sonographic-pathologic correlation in a young man

Maria Adele Marino, MD^{a,*}, Giorgio Ascenti, MD^a, Roberta Cardia, MD^b, Antonio Ieni, MD, PhD^b, Michele R. Colonna, MD^c

^a Department of Biomedical Sciences and Morphologic and Functional Imaging, Policlinico Universitario G. Martino, University of Messina, Messina, Italy

^b Department of Human Pathology in Adult and Developmental Age “Gaetano Barresi”, Section of Pathology, University of Messina, 98123 Messina, Italy

^c Department of Human Pathology of the Adult, the Child and the Adolescent, Section of Plastic Surgery, University of Messina, Messina, Italy

ARTICLE INFO

Article history:

Received 9 September 2019

Revised 8 November 2019

Accepted 17 November 2019

Available online 25 December 2019

Keywords:

Pilomatrixoma

Ultrasound

Imaging

Color-Doppler

Elastography

Malherbe

ABSTRACT

Pilomatrixoma, also known as calcifying epithelioma of Malherbe, is a histological type of benign subcutaneous tumor arising from the cutaneous adnexa. We present our experience and the characteristic findings of a pilomatrixoma of the right thigh in a young healthy man. The lesion had grown slowly in size over the past year and this was the reason for referral to the Department of Plastic Surgery. Ultrasound imaging was performed and, through the use of B-mode, color-Doppler, and elastosonography, the lesion was depicted. The patient was therefore scheduled for surgery.

Pilomatrixomas at times can pose a diagnostic challenge, especially when the location is unusual. Ultrasound and its tools, that is, color-Doppler and elastography, can assist the clinician arising the suspicion of pilomatrixoma.

© 2019 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license.

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Introduction

Pilomatrixoma is a particular and uncommon type benign subcutaneous tumor originating from cutaneous adnexa. Cutaneous adnexal tumors are a large and varied group that commonly are clustered according to their adnexal differenti-

ation based on histologic, ultrastructural, and immunohistochemical analyses: eccrine, apocrine, follicular, and sebaceous [1-3]. Pilomatrixoma was first described in 1880 by Malherbe and Chenantais, hence the name of calcifying epitheliomas of Malherbe, and the terms “pilomatrixoma” or “pilomatrixoma” have been introduced and then used worldwide since 1977 to reflect the more accurate description of a benign ectodermal

Abbreviation: US, ultrasound.

Conflict of interest: The authors declare no conflict of interest concerning the topic of this manuscript.

* Corresponding author.

E-mail address: mariaadele.marino@gmail.com (M.A. Marino).

<https://doi.org/10.1016/j.radcr.2019.11.007>

1930-0433/© 2019 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license. (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)



Fig. 1 – A 15 mm reddish and painful nodule is noted in the upper-inner of the right thigh.

tumor originating from the germinal matrix center of the hair follicle [4].

Pilomatrixoma is the second most common superficial mass encountered in the pediatric age group, second only to epidermoid cysts [3]. It is often a solitary lesion but multiple lesions can occur sometimes and may be associated with myotonic dystrophy, Turner's syndrome, trisomy 9, and Sotos syndrome [5].

Usually, the tumor arises in the head and neck region but other unusual localizations, such as the trunk, have been reported [6]. Only 5% of these lesions occur in the lower limb (5%) [2]. Hmar et al., previously reported a case of pilomatrixoma of the thigh in a 10-year girl [7].

We present our experience and the characteristic findings of a pilomatrixoma of the right thigh in a young man.

Case history

A 20-year-old man presented with an asymptomatic, firm nodule in the 1/3 medial-cranial aspect of the right thigh (Fig. 1). The lesion had grown slowly in size over the past year and this was the reason for referral to the Department of Plastic Surgery. The patient was otherwise a healthy athletic young man. In order to depict the lesion, an ultrasound (US) was performed using a high-frequency linear array transducer (11 MHz; Esaote MyLab E, Milan, Italy). US revealed a well-defined, oval, nodule located superficially in the upper-inner aspect of the right thigh. B-mode US showed a 15 mm solid, oval, hypochoic mass (Fig. 2A). The color-Doppler examination revealed multiple feeding vessels both intralésional and peripheral (Fig. 2B). Strain elasto-sonography was performed revealing the stiffness of the lesion (Fig. 2C). No abnormal lymph nodes were found in the inguinal region. The blood flow pattern on color-Doppler excluded the differential diagnosis of other lesions, such as hemangiomas and vascular malformations. The other differential diagnosis included pilomatrixoma, complicated skin appendage lesion, and sebaceous cyst. Considering the recent increase in size, the pa-

tient was scheduled for surgery. Surgical approach was performed through a lazy S skin incision above the mass, which was isolated, dissected, and removed “en bloc” from the surrounding subcutaneous tissue. The surgical wound was closed with subcutaneous interrupted and intradermal running suture. Healing was uneventful. Gross examination of the removed mass revealed a well-circumscribed, whitish, nodular mass measuring about 2 × 1.5 cm. The whole nodule was fixed in 10% buffered formalin, embedded in paraffin at 56°C; from the corresponding tissue blocks, 4 μ-thick sections were cut and stained by haematoxylin and eosin stain. Histopathological examination showed a dermal encapsulated, partially cystic, proliferation (Fig. 3A) composed by peripheral, monomorphous, basophilic cells (Fig. 3B) and elements with eosinophilic cytoplasm and empty nuclear space, so-called “shadow-cells” (Fig. 3C). Calcifications (Fig. 3D) and occasional multinucleate giant cells were also encountered.

Therefore, the diagnosis of pilomatrixoma with clear resection margins was made.

Discussion

Pilomatrixoma is a benign disease, with an incidence between 0.001% and 0.0031% of all dermato-histopathologic materials submitted for examination [8]. It is a rare disease in adults, while pilomatrixoma may occur in children and young adults, with approximately 45% arising before 21 years. However, a bimodal distribution with a second peak in elderly has been demonstrated [9,10]. Although controversial, the majority of studies report a slight predominance of pilomatrixomas in the female rather than male [11,12]. Pilomatrixoma usually is located in the head and neck region (64%; with the mid face being the most commonly affected area), followed by the upper limb (22%), the trunk (8%), the lower extremity (5%), and other sites (1%). The right side seems to be more prevalent than the left side of the body [2]. Usually the lesions are solitary but multiple pilomatrixomas have been reported and in those cases, the presence of familial or associated conditions should be considered [13].

Pilomatrixomas evolve with time and have been classified into 4 histopathologic stages: early, fully developed, early regressive, and late regressive; based on these criteria our case fits in the fully developed stage [14]. Also, several clinical variants of pilomatrixomas, such as perforating, anetoderma, proliferating, pigmented multiple, and familial have been described. Accurate diagnosis of pilomatrixoma can be made with imaging when they present with typical size and location. On US, the lesion appears as round or ovoid well-defined hyperechoic or heterogeneous solid nodules within the subcutaneous soft tissues. Demonstration of hypo echogenicity, heterogeneity, internal echogenic foci (calcifications) in scattered-dot pattern and hypochoic rim with or without posterior shadowing is highly suspicious for pilomatrixoma with specificity and positive predictability of 95% and 92%, respectively [2,15].

Treatment of choice for pilomatrixomas is surgical excision with histologic confirmation. Most studies reported that com-

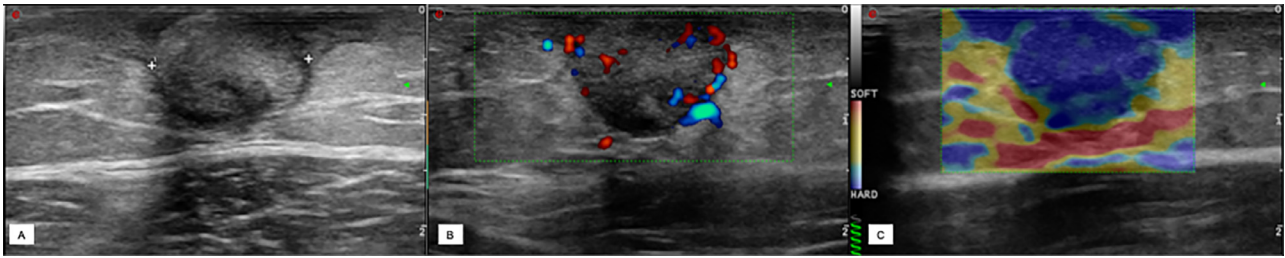


Fig. 2 – Ultrasound imaging of the nodule located superficially in the upper-inner of the right thigh. B-mode ultrasound shows a 15 mm solid, oval, hypoechoic mass (A). The color-Doppler examination revealed multiple feeding vessels both intralesional and peripheral (B). Strain elasto-sonography was performed revealing the stiffness of the lesion (C).

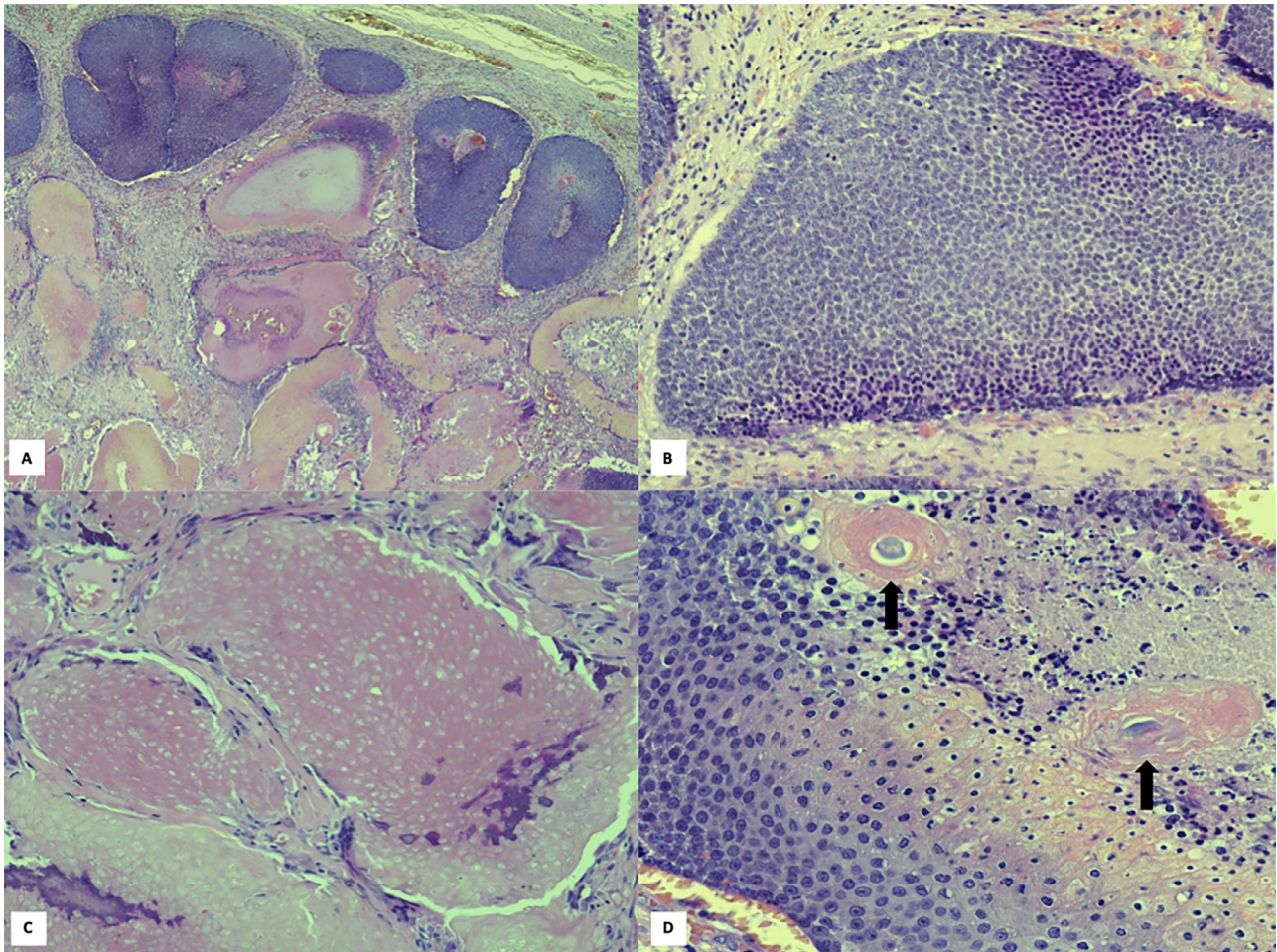


Fig. 3 – Histological examination showed a subcutaneous partly cystic nodular lesion (A, original magnification $\times 10$, Hematoxylin-Eosin), composed by uniform basaloid elements (B, original magnification $\times 20$, Hematoxylin-Eosin) with central keratin debris the acellular material with shadow cells (C, original magnification $\times 20$, Hematoxylin-Eosin) and occasionally were observed also calcifications (black arrows) (D, original magnification $\times 40$, Hematoxylin-Eosin). The diagnosis of pilomatrixoma was made.

plete surgical excision with clear margins is almost always curative. No malignant variants were reported [2].

As this case demonstrates, pilomatrixomas at times can pose a diagnostic challenge, especially when the location is unusual. US and its tools, that is, color-Doppler and elastography, can assist the clinician in arising the suspicion of pilomatrixoma.

Learning points

- Pilomatrixoma, is a particular and uncommon type benign subcutaneous tumors arising from the cutaneous adnexa.
- Pilomatrixoma shows a pattern of high-rigidity at strain-elastography.
- Pilomatrixomas evolve with time and have been classified into 4 histopathologic stages: our case could be included in the 2nd stage “fully developed”.

REFERENCES

- [1] Niwa T, Yoshida T, Doiuchi T, Hiruma T, Kushida K, Mitsuda A, et al. Pilomatrix carcinoma of the axilla: CT and MRI features. *Br J Radiol* 2005;78(927):257–60. <https://doi.org/10.1259/bjr/54676183>.
- [2] Jones CD, Ho W, Robertson BF, Gunn E, Morley S. Pilomatrixoma: A Comprehensive Review of the Literature. *Am J Dermatopathol* 2018;40(9):631–41. <https://doi.org/10.1097/DAD.0000000000001118>.
- [3] Storm CA, Seykora JT. Cutaneous adnexal neoplasms. *Am J Clin Pathol* 2002;118 Suppl:S33–49. <https://doi.org/10.1309/LR16-VURN-JNWC-BOKD>.
- [4] Aherne NJ, Fitzpatrick DA, Gibbons D, Armstrong JG. Pilomatrix carcinoma presenting as an extra axial mass: clinicopathological features. *Diagn Pathol* 2008;3:47. <https://doi.org/10.1186/1746-1596-3-47>.
- [5] Lozzi GP, Soyer HP, Fruehauf J, et al. Giant pilomatrixoma. *Am J Dermatopathol* 2007;29:286–9. <https://doi.org/10.1097/DAD.0b013e318053db45>.
- [6] Beattie G, Tai C, Pinar Karakas S, Cham E, Idowu O, Kim S. Colossal pilomatrixoma. *Ann R Coll Surg Engl* 2018;100(2):e38–40. <https://doi.org/10.1308/rcsann.2017.0196>. Epub 2017 Nov 28.
- [7] Hmar V, Thokchom N, Kshetrimayum S. Recurrent pilomatrixoma of the thigh: an unusual site of presentation.
- [8] Kwon D, Grekov K, Krishnan M, Dyleski R. Characteristics of pilomatrixoma in children: a review of 137 patients. *Int J Pediatr Otorhinolaryngol* 2014;78:1337–41. <https://doi.org/10.1016/j.ijporl.2014.05.023>.
- [9] Yench MW. Head and neck pilomatrixoma in the pediatric age group: a retrospective study and literature review. *Int J Pediatr Otorhinolaryngol* 2001;57:123–8.
- [10] Lan M-Y, Lan M-C, Ho C-Y, et al. Pilomatrixoma of the head and neck: a retrospective review of 179 cases. *Arch Otolaryngol Head Neck Surg* 2003;129:1327–30. <https://doi.org/10.1001/archotol.129.12.1327>.
- [11] Whitemore KR, Cohen M. Imaging and review of a large pre-auricular pilomatrixoma in a child. *World J Radiol* 2012;4:228–30. <https://doi.org/10.4329/wjr.v4.i5.228>.
- [12] Vance A, Seitz WH. Pilomatrixoma of the upper arm in an orthopaedic clinic. *J Shoulder Elbow Surg* 2012;21:e12–15. <https://doi.org/10.1016/j.jse.2012.01.005>.
- [13] Maeda D, Kubo T, Miwa H, Kitamura N, Onoda M, Ohgo M, et al. Multiple pilomatrixomas in a patient with Turner syndrome. *J Dermatol* 2014;41:563–4. <https://doi.org/10.1111/1346-8138.12509>.
- [14] Morgan PR, Accurso B. Clinical pathologic conference case 1: a woman with a lump in her cheek. *Oral Surg Oral Med Oral Pathol Oral Radiol* 2013;115:e34–6.
- [15] Choo HJ, Lee SJ, Lee YH, Lee JH, Oh M, Kim MH, et al. Pilomatrixomas: the diagnostic value of ultrasound. *Skeletal Radiol* 2010;39:243–50. <https://doi.org/10.1007/s00256-009-0678-x>.